OUTCOMES OF CONGENITAL AND INFANTILE CATARACT IN THE UNITED KINGDOM

MELANIE HUN GEE CHAK

SUBMISSION OF THESIS FOR THE DEGREE OF

DOCTOR OF MEDICINE

UNIVERSITY COLLEGE LONDON
UNIVERSITY OF LONDON
2005

ProQuest LLC
789 East Eisenhower Parkway
P.O. Box 1346
Ann Arbor, MI 48106-1346

ABSTRACT

Congenital cataract is a rare disorder but is a priority of Vision 2020, the international programme for the elimination of avoidable blindness, reflecting its treatment potential and the consequences for the child and family if left untreated. As the aetiology of congenital cataract is unknown in the majority of cases with only a minority being preventable, informing secondary and tertiary approaches to prevent visual impairment is currently essential. The quality of life of children with congenital cataract has not previously been reported. This study was a unique opportunity to engage parents and children in the assessment of their quality of life, an important outcome. Equally, data are limited on the impact of amblyopia treatment on the psychological wellbeing of the child with congenital cataract and their families.

The British Congenital Cataract Study (BCCS) cohort comprises a nationally representative group of children who at the time of the present study, have been under management for at least 6 years after diagnosis. 61% of children with bilateral cataracts achieved an acuity of at least 6/18: commonly considered the vision level at which children can be educated at mainstream schools with minimum extra help. The worst median visual acuity (6/48) was recorded in the cataractous eyes of children with unilateral cataract who underwent surgery, suggesting that despite intervention in these children, the outcome is still poor.

Concordance with occlusion is the most important predictor of visual acuity of children with unilateral cataracts. Earlier surgery and concordance with occlusion are associated with better visual acuity and poorer acuity was associated with the presence of a severe cataract and other medical conditions in children with bilateral cataracts. The incidence of postoperative open angle glaucoma was 5.25 cases/100 eyes operated/year. Early age at detection is the most important factor associated with the development of glaucoma after congenital cataract surgery.

The majority of parents found occlusion difficult and almost a third thought their child's behaviour had worsened. Furthermore, a third of the parents thought that the relationship with their child had worsened as a result of occlusion. Despite this, no association was found between occlusion concordance and the child's or parental experience of occlusion and the majority of parents never worried the patches were harmful. The quality of life scores of children with congenital cataract in the present study were comparable to those reported by children with childhood cancers and rheumatological disorders.

This study's findings should inform future parents of children with congenital cataract especially those with unilateral cataract and have implications for resource allocation in terms of schooling and additional help. The findings also emphasise the importance of early detection to enable early surgery and the importance of occlusion to prevent amblyopia. Identification of underlying causes

of difficulties with occlusion may initiate changing emphasis on the management of children with congenital cataract.

Postoperative open angle glaucoma may be the price of successful screening programmes to ensure early detection and treatment of congenital cataract to mitigate against amblyopia. Further work is required to delineate precisely the optimal timing of surgery to balance the benefits of early intervention with potential risks. The finding that the quality of life of children with congenital cataract is comparable with more debilitating and life threatening disorders is unexpected and has implications for ophthalmologists regarding how they view the impact of congenital cataract on the HRQOL of these children.

ACKNOWLEDGEMENTS

I would like to thank first and foremost, Jugnoo Rahi for her constant guidance and support throughout my research training and in the preparation and writing of this thesis. Her unerring enthusiasm, wisdom and humour have been an invaluable source of inspiration. I am grateful to her for all that she has taught me. I also thank David Taylor for his teaching and encouragement, in clinical aspects of my training as well as his advice on the writing of this thesis.

I am grateful to Phillipa Cumberland, for patiently guiding me through the numbers and giving me the confidence to tackle statistics head on. I thank Angie Wade as well, for her statistical advice.

I am indebted to Isabelle Russell-Eggitt, Ken Nischal, Chris Timms, Lynn Speedwell and all the nurses and staff of the ophthalmic unit at Great Ormond Street Hospital for their help in expanding my knowledge of clinical paediatric ophthalmology.

I also thank the children involved in this study and their families and members of the British Congenital Cataract Group without whom this research would not have been possible.

I would also like to thank the Guide Dogs for the Blind Association for funding this study.

Finally, thank you to Michael and Annie Chak and Mark Boden who as always gave me their endless encouragement and support to fulfill my aspirations.

Tables

Figures

1. INTRODUCTION

Congenital cataract accounts for 3-39% of childhood blindness with an estimated 170,000 children being blind globally from this cause (13% of all causes)[1]. Furthermore, the burden of visual impairment and blindness in a child is significantly more than in an adult, as a blind child has many more years of blindness ahead of them with its associated impact on social, education, employment and personal opportunities[2]. Congenital cataract is therefore a priority of Vision 2020 the Right to Sight-the WHO's initiative to reduce the world's burden of blindness[3].

Investigating clinical and patient centred outcomes to inform secondary and tertiary approaches to prevent visual impairment due to congenital cataract is currently essential as the aetiology is unknown in most cases with only a minority being truly preventable.

The British Congenital Cataract Study (BCCS) cohort comprises a nationally representative group of children who were studied previously to determine incidence and aetiology. At the time of the present study, these children had been under management for at least 6 years after diagnosis. This cohort provided an ideal opportunity to investigate these children whose risk factors have been recorded prior to the study of their outcomes.

Previous studies investigating outcomes such as visual acuity and postoperative complications have largely been on selected small case series and may have been analysed using only one eye or have not taken into account multifactorial interactions. Furthermore there have been limited studies on the psychosocial effects of amblyopia treatment and no studies on the health related quality of life of children with congenital cataract. Patient based assessments of the impact of visual impairment on their quality of life-ie vision related quality of life (VRQOL) are now essential [4].

The research described in this thesis was undertaken to provide information on functional clinical outcomes, quality of life and the impact of amblyopia treatment in order to inform better current clinical practice and aid the evaluation of future and existing regimes.

2. BACKGROUND

In this chapter, an overview is presented of issues relevant to the outcomes of congenital cataract with areas of incomplete information being identified. The cohort of children that formed the basis of the work described in this thesis is also described.

2.1 Visual Impairment due to congenital cataract

2.1.1Lens Development

2.1.1.1Normal lens development and growth

The rudimentary eye (optic vesicle) develops as an ectodermal diverticulum from the lateral aspect of the forebrain in the 3rd week of gestation and grows to become the optic vesicle. At the same time the surface ectoderm overlying the optic vesicle thickens to form the developing lens (lens placode) at about 22 days' gestation. The developing lens is induced to grow and develop by the optic vesicle. In turn the lens induces the development of the cornea and stimulates the development of the vitreous body. It is also important for the normal growth of the pigment layer of the retina. How these interactions are brought about remains unknown, but the lens is a vital link in the development of the normal eye[5;6].

In the fetus the lens is almost spherical, is soft and has a reddish tint. By the time it is born its equatorial diameter is two thirds adult size and its anterior-posterior diameter is almost that of an adult. Additional lens fibres are formed and remain throughout life resulting in an increase in size and density of the equatorial diameter of the lens as successive fibres are formed [5].

2.1.1.2 Abnormal lens development

The lens is designed for the transmission and refraction of light and it responds to any insult that disturbs normal development by opacification: a cataract. Cataract pathogenesis has been extensively studied in adults, but less is known about cataractogenesis in children[6-8]. Animal experiments suggest that defects may involve abnormal maturation of lens cells or abnormal interaction of lens fibres. Recently, genetic studies have implicated crystallins (90% of total lens protein, with a key role in transparency), membrane proteins and transcription factors[9-11].

Cataracts vary in shape, position, density and appearance. Due to this variability, the morphology of cataract can be inconsistent even within the same pedigree[12]. Despite this, morphology can still provide important clues as to visual prognosis and possibly aetiology[8]. Cataracts that are dense and nearest to the nodal point have the greatest effect on vision[13], whilst other cataracts may have very little effect on vision such as the anterior polar and lamellar cataract[14].

2.1.1.2.1 Aetiology of congenital cataract

There have been few population-based studies of the aetiology of congenital cataract and a few large case series in which the underlying cause of cataract has been reported. From these, in industrialised countries a cause cannot be determined in 50-56% of bilateral cases and in virtually all unilateral cases[1]. Approximately 20% have a positive family history of isolated cataract with autosomal dominant disease being more common than X linked or autosomal

recessive modes of inheritance. Autosomal dominant inherited disease is a collection of disorders with several different phenotypes. Molecular and genetic studies suggest a considerable phenotypic and genetic heterogeneity with a range of different mutations in several different genes [9;10].

The causes in the remaining 30% are a combination of chromosomal abnormalities such as Down's Syndrome; genetic diseases with lens opacities in association with systemic abnormalities such as Cockayne's syndrome; metabolic disorders like galactosaemia; intrauterine infection like rubella; prematurity and associations with other ocular disorders[1].

2.1.2 Visual development

2.1.2.1 Normal visual development

Vision is not fully developed at birth and its rapid refinement in visual function is paralleled by maturation of mechanisms that control accommodation, smooth pursuit and saccadic eye movements[15]. Visual development is dependent on maturation of the visual system both anatomically and physiologically[5;15].

2.1.2.2 Abnormal visual development

Amblyopia is the term used to describe reduced unilateral or bilateral visual function due to form deprivation and/or binocular interaction in disorders such as anisometropia, strabismus and stimulus deprivation. Amblyopia literally means 'dullness of vision' (from the Greek *amblys*-dull; *ops*-eye) and is important as it is potentially reversible.

Visual experiences in early life play an influential role in the subsequent development and maturation of the visual system. Malleability of the visual system occurs during a period termed the 'critical period' when the 'wiring' of the striate cortex is still pliable and hence vulnerable[15]. This critical period therefore, is the postnatal time span during which the integrity of the visual system needs to be preserved to subsequently permit normal vision in adult life.

Evidence for the critical period comes from both experimental work and clinical observation. Animal experiments on kittens and then infant monkeys showed that depriving an eye of stimulus by monocular suturing of the eyelid resulted in a

reduction of cortical cells serving the closed eye[16;17]. Importantly this phenomenon was not seen in adult animals. Further work showed that the interaction between two eyes is very important in determining maturation of the visual system. Cortical changes due to monocular closure may not be caused purely by disuse but also depend on the status of the other eye[18]. Competition between eyes results in the eye with any visual advantage dominating the visual cortex[16]. If monkeys with monocular eyelid closure underwent closure of the fixating eye, then the vision improved in the originally closed eye demonstrating the potential reversibility of the effects of visual deprivation[17].

This experimental work has had important implications for the understanding and treatment of all forms of amblyopia. Valid extrapolation of the animal experiments to humans is borne out by clinical observations and studies, although the time of the critical period differs. In humans, there is a brief initial period postnatally of insusceptibility to a degraded image[19]. This is followed by a period which lasts up until a few months of age when the developing visual system is readily degraded by visual deprivation. Between the ages of 6 to 18 months there is the most rapid rate of decline in sensitivity to deprivation with a further gradual decline until the age of 9 or 10. By this time the child's visual system is no longer affected by degraded visual input and equally is no longer readily reversible by treatment[20]. However, recent work suggests that some degree of plasticity can exist throughout life. Any deprivation effect is potentially reversible and will be less profound if vision is restored before the critical period ends[16;21;22].

2.1.2.3 Abnormal visual development due to congenital cataract

In eyes with congenital cataract that are otherwise healthy (the majority), visual loss is primarily the result of amblyopia. This arises in a number of ways. Firstly, and most profoundly, stimulus/form deprivation during the critical period of visual development caused by untreated cataracts. Secondly, competition between the eyes, which is especially important in unilateral cases and can have a bearing on asymmetrical bilateral cases. Thirdly, inadequate correction of refractive error and finally stimulus/form deprivation in postoperative complications such as posterior capsular opacity and postoperative astigmatism[23;24]. Other mechanisms such as those that occur in postoperative complications like postoperative open angle glaucoma and retinal detachment can also cause visual impairment directly and by causing amblyopia.

2.1.3 Impact of visual impairment due to congenital cataract

2.1.3.1 Impact of visual impairment due to congenital cataract on the individual

Visual impairment and blindness in childhood, irrespective of its cause has implications for the child throughout life. Furthermore, the burden of visual impairment and blindness in a child is significantly more than in an adult, as a blind child has many more years of blindness ahead of them with its impact on social, education, employment and personal opportunities[2].

2.1.3.2 Impact of visual impairment due to congenital cataract on the population

Although prevalence and incidence rates of congenital cataract vary between industrialised and developing nations, it remains a major cause of preventable blindness worldwide and has therefore been targeted as one of the main priorities of the World Health Organisation's VISION 2020 programme 'The Right to Sight'[3]. It accounts for 3-39% of childhood blindness (as defined by the World Health Organisation (WHO) classification, (Table1) with an estimated 170,000 children being blind globally from this cause (13% of all causes)[1].

Table 1: WHO Classification of visual impairment

Level of visual impairment	Category of vision	Visual acuity in better eye with optical correction
Sight if visual acuity <6/7.5	Normal vision	6/18 or better
Visual impairment	Low vision	<6/18 to 6/60
Severe visual impairment	Low vision	<6/60 to 6/30
Blind	Blindness	<3/60 to no light perception or visual field ≤10 degrees around central fixation.

A recent study of children diagnosed with severe visual impairment or blindness in one year in the UK (2000-2001) found that 21/439(5%) cases were due to cataract[25]. Nearly all of these children had treatable disease in which severe visual impairment or blindness could have been avoided[25].

The estimated global cost of childhood blindness due to cataract in terms of care and lost productivity is between 180 million and 10,000 million US dollars. The majority of this cost is by children in high income countries where life expectancy and productivity is greater than in low income nations[26-28].

2.1.3.3 Frequency of congenital cataract

Variation in the reported prevalence of congenital cataract is due to the different methodology, age groups and case definitions in different studies, as well as true differences in the populations. The prevalence at birth of bilateral cataract in industrialised nations is 1.2 to 3.0/10,000 births (Table 2) and given a birth rate of 2% (20,000/million), 4 children/million total population/year will be born with bilateral cataract in industrialised nations. Rates for developing countries is likely to be approximately 10/million/total population/year due to greater exposure to infectious agents such as rubella and consanguinuity being more common[29;30].

In industrialised nations, approximately 2 to 4 infants out of every 10,000 born will be diagnosed with cataract by their first birthday with a further 1 child being diagnosed by the age of 15 years[31;32].

Table 2: Prevalence of congenital cataract

Country	Method of study	Year of study	Age examined	Prevalence/ 10,000
Industrialised countries				
USA[33]	National Surveillance	1988-91	Neonates	1.2
France[34]	National Surveillance	1979-88	Birth	2.2
Denmark[35]	Population based prevalence	1984	5-13 years	2.3
UK[36]	Cohort	1984	Birth 2-5years	3 3
Sweden[37]	Population based prevalence	1971	4 years	4
UK[38]	Cohort	1970	10 years	3.3
Non- industrialised countries				
China[39]	Population based prevalence	1987	0-18	3.6
Malawi[40]	Population based prevalence	1983	<6	3.7
Nepal[41]	Population based prevalence	1980	<10	1.7

2.1.4 Prevention of visual impairment due to congenital cataract in the United Kingdom

Primary, secondary and tertiary strategies are aimed at preventing visual impairment due to congenital and infantile cataract and are discussed in detail below. Primary strategies aim to prevent exposure to risk factors that lead to congenital cataract. Secondary prevention focuses on stopping or slowing the progression of disease or on preventing visual impairment occurring due to established disease such as screening and detection for early diagnosis, treatment and follow up. Finally, tertiary prevention is directed at managing and rehabilitating children with established visual impairment despite treatment, for example using low visual aids and additional educational support.

2.1.4.1 Primary strategies

It is generally advised that preconceptual genetic counselling for families with a history of hereditary disease should be offered consultation with an ophthalmologist. Rubella immunisation programmes are implemented and maintained in most industralised countries[42]. Public health education programmes should also be instigated to increase awareness and understanding of hereditary eye disease and to promote avoidance of known teratogens[26;29].

2.1.4.2 Secondary strategies

In the UK screening for ophthalmic disorders are undertaken within a broader context of a national programme of child health surveillance. The Children's Sub-

Group of the National Screening Committee (NSC) of the UK reviewed completed studies and work under way as well as eliciting expert opinion to make national recommendations on the screening for visual deficits and ophthalmic disorders[42]. The National screening committee recognises that early treatment is essential to prevent amblyopia *(refer to section 2.1.2 Visual Development, page 21)* and that this is particularly important in unilateral cataract (*refer to section 2.2.2 Management of congenital cataract by the laterality of the cataract, page 34*). However early treatment is only possible if the child with congenital cataract is detected early and referred promptly[43;44].

The National Screening Committee recommendations are that all newborns should be screened for media opacities, comprising examination of the pupillary red reflex, inspection of the eyes and inquiries about the visual behaviour of newborn children. A repeat examination not later than 6 weeks for cataract and other eye anomalies is recommended[42]. These are consistent with recent guidelines in the USA[45]. However, problems have been identified in the training and supervision of the screening paediatricians in the UK and the need for improvements in their training are recognised[46;47]. The NSC also recommends that children with ophthalmic disorders associated with other major disabilities, should be examined by an ophthalmologist and an orthoptist and children with other neurodevelopmental disorders. In particular those with hearing loss should have an expert eye opinion as a matter of routine[42].

Other reports recommend routine examination of children with a known family history of hereditary cataracts to provide prompt detection [26;29]. Public health programmes to promote parental understanding of the importance of early detection and treatment of congenital cataract are also advocated [26;29].

2.1.4.3 Tertiary strategies

It is considered good practice to provide follow up of cases to detect and treat postoperative complications, especially in the long-term for conditions such as glaucoma. In children with visual impairment, assessment and provision of low vision appliances as well as provision of educational support should also be made available. For cases that present late, where appropriate, surgical treatment should be provided in order to provide cosmetic improvement or navigational vision[42].

2.2 Management and outcomes of congenital cataract

Visually significant congenital cataract must be removed surgically, or in some circumstances conservatively managed before subsequent removal. In untreated cataracts, some are sufficiently mild that they are little changed by time, such as lamellar cataracts[8]. The risks of surgery and amblyopia treatment mean that it is inadvisable to treat children with congenital cataract unless it is certain that without treatment the child's vision will be insufficient for mainstream schooling and social activities. However, if the vision is sufficient for this purpose, active management can be postponed until the child is older, when surgery maybe technically easier and optical correction less difficult to achieve[8].

Whilst outcomes of congenital cataract such as visual acuity and postoperative complications have been extensively reported, previous studies have been largely based on small and/or selected case series of patients and have been complicated by factors such as the analysis of unilateral and bilateral cases together, differing surgical techniques used and short follow up. There have been no population based studies of outcomes of congenital/infantile cataract and only 3 randomised controlled trials, all in India, of treatment of children with congenital/infantile cataract[48-50].

2.2.1 Measurement of visual function in children

As the primary aim of the clinical management of children with congenital cataract is to improve their vision, measures of visual function are important in the assessment and management of children with congenital cataract. These measures include visual acuity, visual fields, contrast sensitivity and binocularity and are all relevant to overall function. 'Normal' ranges have been identified for some of these[51]. Difficulties and thus inaccuracies in testing, especially of very young children, are related to their cognition and immature language skills.

Distance visual acuity remains the most frequently measured visual function in clinical practice. This is probably due to the ease and speed with which its assessment can be executed compared to other modalities, its good repeatability and that it gives a measure of function. It is therefore a regular part of the routine assessment of children in ophthalmic clinics.

A variety of optotype tests are available for testing distance visual acuity in children. They include the use of pictures (eg Kays Pictures), matching tests (eg Sonksen Silver test, Sheridan Gardner test) or forced choice preferential looking techniques (eg Cardiff cards). Isolated optotypes overestimate visual acuity in amblyopia (the main cause of visual loss in children with congenital cataract), therefore it is important that crowding boxes or full charts be used[52]. Many of these tests can be modified to include elements of crowding to improve detection of visual loss from amblyopia (eg HOTV optotypes, crowded Kay picture test). In

adults and children over 6 years, the logMAR Early Treatment Diabetic Retinopathy Study protocol has been successfully used and is reliable and repeatable[53;54], but in younger children there is no commonly accepted testing method[55].

Most of these tests require a distraction free environment and the child to concentrate and cooperate. In experienced hands however, they provide good measures of visual acuity. Reliability studies indicate that children are comparable to adults[56].

2.2.2 Management of congenital cataract by the laterality of the cataract

Issues surrounding the management of children with unilateral and bilateral cataracts in terms of outcomes and their factors differ primarily because they are different amblyogenic situations. Furthermore, the prognosis of the non-cataractous eyes of children with unilateral cataracts is extremely good, contrasting with the poor prognosis of the cataractous eyes, especially if the child is not treated. Therefore, children with bilateral and unilateral cataracts are managed differently with regards to timing of surgery and postoperative management and as such should be studied as two distinct disease entities.

Children with bilateral cataracts are reported to have a better visual outcome compared to unilateral cataracts[57-59]. This is because bilateral stimulus deprivation prolongs the critical period and a competitive interaction between the eyes is reduced[60]. The critical period in bilateral cataracts is uncertain, and some propose that the first 6 weeks of life represents a latent period for binocular development[61], but certainly by 4 months of age the child will be showing signs of visual deprivation if the cataracts are not removed[20]. The first eye that is operated on in bilateral cataracts usually has the better outcome, especially if operated on early[21] and in asymmetrical cataracts, the more severely affected eye develops the worse vision[8].

In unilateral cataract the imbalance between the eyes results in the cataractous eye being at a disadvantage in competing for dominance in the visual cortex and

it will become amblyopic if not treated. The general consensus for best possible outcomes for children with unilateral cataracts is early surgery (before 6 weeks of age) [62-64] followed by aggressive occlusion regimes[65] and timely correction of aphakia[66]. Nevertheless, historically, unilateral cataract outcomes have been poor[67]. Since the early 1980s however, advances in surgical technique have allowed safer and earlier surgery (*refer to section 2.2.3 Surgical techniques, page 36*). This, coupled with better assessment of the visual acuity (VA) with preferential looking techniques allowing more accurate assessment of very young infants and titration of treatment[65;68], have resulted in good VA in some of these children.

However, current debate on the management of unilateral congenital cataract centres on whether the benefits outweigh the risks in the treatment of what is essentially a non-blinding condition. The management of unilateral cataracts is intensive with high involvement of the family in postoperative occlusion regimes (causing possible psychological trauma) as well as multiple clinic visits (*refer to section 2.2.5.2 Occlusion and penalisation, page 47*) and the risk of postoperative complications. Also, when children present late with unilateral cataracts there is the additional quandary of whether the cataract was there during the critical period and it is often difficult to ascertain whether borderline cases are truly visually significant[69]. Advocates of treatment argue however, that treating the cataractous eyes allows a 'spare' eye and binocular vision may be achieved with a wider field of view[69].

2.2.3 Surgical techniques

Cataract surgery aims to restore a clear visual pathway. Simple lens aspiration (no vitrectomy or capsulectomy), popularised in the 1960s[70], was routinely performed and the long term follow up of this procedure suggested that the complication rate was low[8;71]. However, amblyopia induced by posterior capsular opacity (PCO) remained a problem until posterior capsulotomy or capsulectomy either at the primary procedure or later as a secondary surgical or laser intervention overcame this problem. As vitrectomy machines became more common in the 1970s, they were used to prevent the problems of PCO and vitreous strands[8], resulting in lensectomy with vitrectomy becoming the routine procedure as it removed the entire capsule.

More recently in children older than 2 years, lens aspiration with a posterior capsulorhexis (limbal or pars plana)[72] and a limited anterior vitrectomy has become the treatment of choice. This is because it conserves the capsule allowing placement of an intraocular lens as a primary or secondary procedure,[73] with medium to longer-term outcome data becoming available[8;69;74-85]. In children younger than 2 years old, although widely practiced, the case for the longterm safety of this procedure is still not made (*refer to section 2.2.5.1 Refractive Correction, page 43*).

2.2.4 Complications of cataract surgery

Complications following cataract surgery are important as they reduce potential visual function. They may also have an important impact on the child and parents' general well being with disruptions due to repeated operations and procedures and numerous hospital appointments as well as worry of the future. Post operative open angle glaucoma in particular frequently has a devastating effect on vision, being hard to detect until there is permanent visual loss. It is also difficult to treat or control, making the need to identify factors associated with this disorder important.

The complications of infantile cataract surgery have been reported by a number of authors, but their incidence rates have rarely been reported and interpretation of trends requires understanding of advancement of surgical technique and improvements in technology and equipment. Thus, reports of newer techniques such as Intraocular lens (IOL) implantation, will have shorter follow up and may have misleadingly low complication rates[86].

2.2.4.1 Postoperative glaucoma

Postoperative closed angle glaucoma is a well documented complication of paediatric cataract surgery[87]. It occurs either due to a fibrinous membrane forming over the pupil or prolapse of the vitreous forward[12]. It tends to occur early, is relatively easy to diagnose and is decreasing in frequently due to primary vitrectomy and postoperative steroidal and cyclopegic drugs administration becoming a routine part of the cataract extraction[88;89].

In comparison, postoperative open angle glaucoma is emerging as potentially the most important visually disabling consequence of cataract surgery and alarmingly, may be one of the commonest[12]. Proportion varies from 6 to 32%, depending on the series and length of follow up[14;85;86;88-93]. On examination of these children, the angle is open on gonioscopy and may have micro PAS and abnormal pigment deposition[94]. It is insidious, can be difficult to detect, may occur many years after surgery[88-90;90;95;96] and it is well recognised that all aphakic children should be considered at risk for the remainder of their lives[8].

Postulated possible aetiology of open angle glaucoma includes genetic predisposition[86;88], high dose steroids and/or vitreous factors altering the trabecular meshwork development or maturation of the angle[94]. To date, from case series, the most likely factors predisposing to postoperative open angle glaucoma appear to be intra-ocular surgery at less than 16 months of age, poor pupil dilatation, microcornea < 9mm, and the need for secondary

surgery[85;88;91;93;97;98]. There are however, no population based studies looking at risk factors.

Differing cataract surgery technique with variable amounts of manipulation and removal of the vitreous may also possibly influence the onset of glaucoma[89]. The shorter follow up for the newer lensectomy-vitrectomy technique versus older techniques makes direct comparison difficult especially as postoperative open angle glaucoma can occur many years after surgery. Consequently, the length of follow up is an important issue in the interpretation of reports[8;85].

Some have postulated that the presence of an IOL helps prevent glaucoma. One review of the literature reported no glaucoma in over 1000 eyes operated on for congenital cataract with implantation of an IOL, however most children were over 2 years old and microphthalmic eyes were excluded[99]. This conflicts with the reports of others in which glaucoma occurred more frequently in eyes with an IOL[100-103].

Treatment of postoperative open angle glaucoma is extremely difficult and requires lifelong intervention. Medical treatment inevitably fails, leading to surgery and/or laser treatment[104]. Cyclodiode can sometimes provide long-term control, but the majority of patients will require further laser within the year and virtually all need supplementary medical treatment[105]. Surgery has also been reported as disappointing. Trabeculectomy, even with mitomycin C, has high failure

rates[104;106], with one study with the longest follow up (mean=18 months) reporting 100% failure[107]. Drainage procedures appear to be more promising and adjunctive antifibrosis therapy appears essential for success despite the greater risk of over drainage[104].

There is therefore a pressing need to identify risk factors and predictors of postoperative open angle glaucoma in order to inform preventative strategies as currently, treatment modalities have limited success[104;106].

2.2.4.2 Retinal detachment

Postoperative retinal detachment has been reported in case series to occur in 1% to 10% of aphakic eyes[8;85;108] and mostly occurs many years after surgery. The risk and natural history of retinal detachment in pseudophakic eyes, subjected to different surgical techniques, has not been extensively studied although it is postulated that it is associated with vitreous traction especially in aspiration techniques[108;109].

2.2.4.3 Endophthalmitis

Endophthalmitis is one of the most serious complications following cataract surgery as it has devastating consequences on vision. Risk factors include nasolacrimal duct obstruction, periorbital eczema and upper respiratory tract infection[110]. In a survey of over 500 consultants, endophthalmitis was reported in 7 of 10,000 children undergoing surgery for cataract or glaucoma[71].

2.2.4.4 Posterior capsular opacity and secondary membrane formation

Posterior capsule opacity (PCO) is a well recognised complication of cataract surgery in children, with improved visual outcomes during the 1980s being attributed, in part, to the elimination of deprivation amblyopia from PCO[111] through primary surgical capsulectomy or secondary Neodymium:YAG (Nd:YAG) laser capsulotomy[112]. The management of PCO has taken a new direction with the desire to maintain some of the posterior capsule for IOL implantation. As the presence of an IOL *per se* increases the risk of PCO, primary capsulotomy in IOL implantation is especially advocated[102;112;113;114], but recent reports indicate that secondary membrane formation or PCO is common despite this[103;115].Younger age (≤1 year old), the presence of a capsule, an IOL and absence of vitrectomy, all independently appear to increase the risk of secondary membrane formation[111;116;117], whereas the type of IOL material does not appear to make a difference[117]. New techniques in placement of the lens have appeared to reduce PCO development[8;118].

2.2.4.5 Significant postoperative inflammation

Severe post-operative intra-ocular inflammation is recognised and is often difficult to control in young children[119;120;121]. This is probably related to a shallower, less maintainable, anterior chamber making these small eyes prone to iris damage and uveitis[122]. Thankfully however this is decreasing in frequency with rigorous and intensive postoperative regimes.

2.2.4.6 Sympathetic Ophthalmia

This is a rare complication and is rarely reported in the literature but is severe enough to warrant the preoperative discussion of the possibility[123].

2.2.4.7 Other postoperative complications

Most other complications are usually visually insignificant, such as pupillary damage, heterochromia and transient retinal haemorrhage[8;124;125]. Vitreous strands or wicks are not unusual and this is increased with postoperative crying inducing vitreous prolapse[8].

Cystoid macular oedema does not appear to occur in the early period following cataract surgery. It may be a significant cause of visual morbidity longterm but it is rare probably due to good vasculature in children[126]. Some postulate that the disturbance of the vitreous (such as in vitrectomy and PCO management) may lead to cystoid macular oedema[127].

2.2.5 Prevention and treatment of amblyopia

Amblyopia may be caused directly by the opacity in the visual axis, strabismus, as well as blurring of the image from uncorrected aphakia. Therefore the prevention and treatment of amblyopia in congenital cataract is threefold: removal of the cataract, correction of the aphakia in order to produce a sharp retinal image preventing stimulus deprivation and occlusion if there is unequal competition between the eyes.

2.2.5.1 Refractive Correction

Once the lens has been removed, rendering the child aphakic, the refractive error must be corrected to prevent amblyopia. There are four possible modalities: contact lenses, glasses, intraocular lenses or epikeratophakia.

Contact lenses

Contact lenses (CLs) allow accurate and easily altered refractive correction as the eye elongates with age and are well tolerated by children and their parents with 85% in one series successfully using them: most started CL wearing in the first months of life[128]. The difficulties, however, of contact lens use in younger children may be under reported. In a recent survey, 123 caregivers reported contact lenses to be reliable, easy to use and well tolerated by their children. Furthermore, 87.8% of caregivers preferred CLs to intraocular lenses, given that the visual acuity would be the same and the children would need some sort of additional refractive correction with IOLs anyway[129]. Self CL use in older children

can be taught to those as young as 11 years, who have been reported to successfully handle lenses after only 6 months' training[95].

Nevertheless, there are potential difficulties, as lenses are easily lost and infectious keratitis, hypoxic corneal ulceration and vascularisation can occur[122]. Poor concordance is also a recognised problem with contact lenses, but this is generally due to amblyopia rather than difficulties with the CL itself[130]. Some of these problems could be avoided by fitting permanent wear silicone lenses which are safe up until the age of 3 (when significant deposit buildup becomes a problem) as long as there is adequate support services from optometrist, orthoptists and emergency ophthalmologists[131].

Glasses

Glasses are safe to use and have the additional advantage of magnifying the image, thereby improving acuity. They may also confer a cosmetic advantage by magnifying microphthalmic eyes. However, glasses also have limitations. They are of limited use in unilateral aphakia due to aneisokonia and are cosmetically poor[8]. They also have problems with image distortion, deficiencies in peripheral vision, difficulties with fitting and problems in varying vertex distances in an active child, all of which can affect visual outcome[74;75;95;129;132].

Intraocular lenses (IOL)

IOL implantation has become common and has become the standard therapy in treatment of children >2 years old with cataract[74;75;95;132;133]. An IOL offers the potential for refractive correction which better approximates the natural crystalline lens, and with which at least partial optical correction is always in place and it is argued that this is a less amblyogenic situation, particularly as contact lenses can be damaged or lost[100;101]. Advocates argue that the visual outcomes with IOLs are comparable with those with aphakia corrected with contact lenses or glasses. Much of the early impetus for IOL implantation in young children was the difficulty of managing amblyopia in those with unilateral cataract.[69;100;101]

A survey of ophthalmologists attending the American Association for Pediatric Ophthalmology and Strabismus in 2001 showed that IOL implantation is being performed at a younger age compared to 8 years ago, with an overall median age at implantation of 0.5 months and 99.3% of surgeons implanting at less than 2 years of age[134]. However, the use of intraocular lenses in infancy is controversial with only short term data on small case series of selected children[74;75;100;102;113;119-121;135-141]. IOLs may introduce additional risks such as repeat surgery for repositioning of lenses or IOL induced uveitis. The IOL must last the lifetime of a child, which is considerably longer than the experience of the use of PMMA lenses in adults. Similarly, acrylic lenses are gaining in popularity but follow up for these is even shorter[142;143]. Bio-compatibility, sizing and design issues are also important but have been investigated only in post-mortem eyes[144].

Finally, predicting the final refractive error in children, especially in children less than 2 years old is difficult because although the mean refraction in aphakia declines logarithmically (myopic shift) from infancy to 20 years[145], with a theoretical rate constant [101;146;147], the variance of the rate is large and markedly so in younger eyes[148;149]. Given the inaccuracy of the IOLs in correcting refractive error, additional refractive correction may be necessary anyway for these children.

Secondary IOL implantation is possible in eyes which have enough posterior capsule to support the lens and it is usually implanted in the sulcus[121]although in selected cases can also be placed in the bag of the capsule itself[150].

Epikeratophakia

This procedure was once in vogue but has now largely been abandoned due to unpredictable results in visual acuity and refractive outcome, prolonged graft haziness and the significant risk of corneal scarring in failed procedures[151].

2.2.5.2 Occlusion and penalisation

The effectiveness of occlusion therapy in congenital cataract has never been addressed in a randomised control study due to clinicians' concerns that it would be unethical to have a non- treatment group[152]. The only randomised controlled study of amblyopia treatment reported on children with mild to moderate amblyopia (none with congenital cataract) found that treatment is worth while in children with the poorest acuity only (≤6/18)[153]. Most available studies have concentrated on other forms of amblyopia (e.g. strabismus or anisometropia) and it is unknown if findings from these can be extrapolated to children with congenital cataract.

Nevertheless, there is agreement that poor concordance is associated with poorer visual outcomes in children with congenital cataracts[8;154;155] and occlusion of the phakic eye in unilateral cataract is as essential as early surgery and optical treatment[8;156]. Poor concordance is associated with social deprivation probably because of reduced access to healthcare[157] and may be improved by better parental education[158;159].

A wide variety of aggressive occlusion regimes have been used, especially in unilateral aphakia ranging from at least 50% to nearly full time occlusion of the fellow eye[12;66;67]. A less intense regime of occlusion appears to improve binocular vision, but does not appear to decrease the visual acuity[160]. These children have dense amblyopia, and the findings of a recent study (of severe amblyopia, but not

congenital cataract) suggested that 6 hours is just as effective as full time occlusion in improving visual acuity[161]. Irrespective of regime, occlusion therapy should continue until visual maturity[162].

Although occlusion aims to improve the vision of the aphakic eye, it can compromise the visual acuity (confirmed on electrophysiology studies) of the good eye in unilateral cataracts.[163;164]. This loss is not apparent in children with untreated uniocular visual impairment and may therefore be an iatrogenic effect of occlusion[164].

Despite the prolonged occlusion treatment that is used in many children with cataract, invariably in those with unilateral disease, the neuro-developmental and psychological consequences have received limited attention. The literature suggests that children with monocular cataract are similar to their siblings in terms of their development and behaviour at ages 1 to 16 years[165], and that the early mental development (Bayley Scales) of children having surgery aged <8 weeks is better than those having surgery later[166]. A survey of caregivers of children with congenital cataract reported that occlusion was more stressful than contact lens insertion and removal and cataract surgery[129]. There were no differences between the age groups (less than 2 years and older than 2 years), in stress or difficulty levels[129].

A recent study used a newly developed questionnaire to assess the acceptability of occlusion and atropine treatment of 364 children, and found that both modalities of treatment were acceptable[167], whilst a qualitative study reported that parents reported distress with occlusion and this affected concordance[168]. Neither of these study groups included children with congenital cataract. Research on the psychological impact of occlusion following cataract surgery, which is invariably intense, would be of interest.

Pharmacological penalisation is an established method of treating anisometropia and strabismic amblyopia[153;169] and may therefore be effective in bilateral aphakia. It is ineffective in amblyopia secondary to unilateral aphakia due to the severity of the amblyopia, the absence of accommodation in the aphakic or pseudophakic eye and the disparity between the eyes being so large[12].

2.2.6 Quality of life

2.2.6.1 What is quality of life?

The importance of quality of life (QOL) has been recognised since ancient times. Aristotle in his Nicomachean ethics[170] relates quality of life to happiness and the philosophical approach that QOL is how one rises to challenges and copes with adversity:

'When a man bears patiently a number of heavy disasters, not because he does not feel them but because he has a high and generous nature, his nobility shines through. And if, as we said, the quality of life is determined by its activities, no man who is truly happy can become miserable; because he will never do things that are hateful and mean'.

In current times, QOL is an essential assessment of modern life. The British government now recognises 15 indicators of QOL, identified by the general public as well as business and environmental groups which are assessed annually and range from violent crime rates to air quality[171]. QOL however means different things to different people and is part tempered by the use of the term loosely in everyday language. Definitions and ideals depend on specific cultural, social, spiritual and social circumstances. Specifically, the social and psychological approach emphasises the subjective nature of QOL, questioning who should best judge QOL, and thus defining the benchmark for setting standards.

2.2.6.2 Health related quality of life (HRQOL)

The notion of HRQOL stems from the aspiration that the application of healthcare is to improve a patient's life and embodies all of the factors discussed earlier. With improvements in medical care, survival is no longer the main objective in determining the success of healthcare provision. HRQOL refers to the impact that illness and its treatment have on a patient's quality of life. There is a consensus that it is multidimensional; encompassing the physical, social and psychological effects of treatment. However, definitions have varied, with some based on function or disabilities and others on the degree of (mis)match between aspirations and experiences[172-174]. Importantly there is increasing emphasis on the subjective nature of HRQOL. Thus it is recognised that HRQOL will vary between individuals and at different time points and people with different expectations will report a different quality of life even if they have the same clinical condition, interventions and/or functional outcomes.

2.2.6.3 Why measure HRQOL?

The WHO definition of health in 1958 was 'a state of complete physical, mental and social well-being and not merely the absence of disease and infirmity'[174a]. The definition of health is further qualified as: 'The individual's perception of their position in life and the context of culture and value systems in which they live and in relation to their other goals, expectations and standards'.

A medical condition, especially if chronic, may have an impact on many aspects of life in addition to the specific demands of the illness such as hospital visits and time off school. Thus assessing HRQOL adds a different dimension to assessing the outcome of medical care than clinical information. Importantly, in ophthalmology, where illness is rarely associated with death, treatment is primarily aimed at improving functional vision and thereby indirectly, HRQOL. In addition, the use of HRQOL measures in clinical practice encourages a focus on the patient and not the disease as well as providing a way of increasing patient involvement in decision making and self- care. HRQOL can be used to compare alternative treatments and justify and prioritise expenditure of budgets. Assessment of HRQOL can also facilitate communication between patient and doctor, as well between doctors in shared care situations[175].

However, despite the importance and usefulness of HRQOL measures they rarely actually contribute to clinical decisions[176]. This is partly because there are a number of difficulties in assessing HRQOL (as discussed below) and there are few studies on the implementation of new schemes and audits on their effectiveness[175]. Nevertheless if measuring a child's HRQOL became as routine as measuring the visual acuity there would be a beneficial impact on clinical management.

2.2.6.4 Generic versus Specific HRQOL measures

Any QOL measure should be valid (measures what it was intended to measure), reliable (consistently yields the same results in repeated applications on an unchanging population or phenomenon), responsive (detects changes in QOL associated with treatment or illness) and assess a wide range of behaviours and activities[177]. Both generic and specific HRQOL measures exist. Generic measures have breadth of application, having been designed to capture all aspects of the QOL. Thus they allow comparison of sick and healthy populations as well as across different disease groups. Generic measures tend to have been developed on large samples and are therefore potentially more statistically robust. Although the main criticism of generic measures is that they can lack the sensitivity and specificity of disease-specific measures, in certain populations and diseases, some generic tools are just as reliable and specific as specific tools[178]. In the main though, specific measures are considered better at evaluating the impact of different interventions and treatments within a specific patient population. Specific measures may explore a single domain in greater depth than a corresponding generic measure but can also tend to have a more narrow focus.

2.2.6.5 Problems with measuring HRQOL

People have different expectations which for a given individual will change over time as their disease and any interventions develop, ie a 'response shift'. The point at which the HRQOL is measured may occur at different points on their illness trajectory and it is impossible to ascertain where that exact point is[179]. At

present measures are unable to distinguish between changes in the experience of disease and changes in expectations of health.

The act of measuring HRQOL in a clinical setting may generate the expectation that the clinician will be able to improve the situation, which is not always possible and can result in patient harm. In addition, the sheer breadth of concepts addressed in examination of HRQOL may identify factors that are outside the remit of usual medical care. In the US, pressure groups have opposed the over 'medicalisation' of life and interference in aspects of patients lives that should not concern the clinician[175].

2.2.6.6 Measuring HRQOL in children

The 1989 Convention on the Rights of the Child stressed the rights of the child to adequate circumstances for physical, mental, spiritual, moral and social development. (Centre for Human Rights, United Nations. *Convention on the rights of the child.* Geneva:United Nations, 1989). Moreover, a child has the right to express their opinion freely and have that opinion taken into account in healthcare as in other domains[180].

Measurement of QOL in paediatrics is therefore theoretically as important as in adults, however it presents greater challenges. Any measure needs to assess the child's view of their own health and functional status and to be able to accurately gauge the child's conceptual and developmental viewpoint of their quality of life, whilst taking into account the normal physical, emotional and social

developmental changes[174]. Children's concept of health changes as they mature[181] and any QOL measure must be sensitive to these normal changes in childhood. Older and more cognitively mature children conceptualise illness in terms of specific symptoms and disease rather than a global non-specific understanding and are more aware of psychological, social and emotional implications of illness compared to younger children[182]. Furthermore, a child's understanding of illness may be related to either or both their experience of a disease and their cognitive development[174].

Proxies, including medical professionals, carers or relatives answer questions on behalf of the child in situations where the child is unable to answer questions on QOL themselves. Most commonly, parental proxies are used, however there is now a body of evidence to support the use of child completed QOL measure in parallel with parent rating rather than relying on proxy reports alone. This is because parental assessment of their child's HRQOL may be inaccurate. Agreement between proxy and child is influenced by the relationship between them and parental distress about their child's disorder may influence their assessment of HRQOL[183]. It may also be difficult for parents to appreciate factors that are most important to their child and overall, children are less concerned with basic functional tasks. Specifically with regards to HRQOL, children do not share adult's views about cause, aetiology and treatment of illness and may interpret questions differently as well as adopt a different time perspective regarding the course of a disease[177;184]. On this basis, adults may not be able to rate a child's

QOL well because their priorities and framework about life differ so greatly[173]. By contrast, the child will have limitations on their abilities to use rating scales, understand language and complete questions of the type used in adult tools and so completion may be compromised by their cognitive development.

2.2.6.7 Measuring HRQOL in ophthalmology

Clinical outcomes, especially visual acuity, have been the primary variables of interest in most epidemiological studies of the blind and visually impaired and are also the main outcome in clinical intervention trials. Visual acuity is also used to define the legal requirements for driving, the eligibility for blind and partially sighted registration and to guide clinicians on aspects of care such as surgery. The changing health care system now places stronger emphasis on patient led assessment of health care outcomes; therefore solely physical measures of visual impairment are now inadequate descriptors of visual function. Patient based assessments of the impact of visual impairment on their quality of life-i.e. vision related quality of life (VRQOL) are now essential [4]. This is reflected in the Department of Health's "Patient empowerment at the heart of the government's plan to modernise the health service" (Press Release, DOH 18/04/2000).
To date, all VRQOL instruments have been developed for adults and most are primarily concerned with visual function in different diseases with limited exploration of psychosocial issues[184-189]. They are also limited in their ability to capture the broader impact of being unable to perform tasks and the importance of visual impairment[190]. They have, however, been used in studies on a variety of

ophthalmic conditions including age related macular degeneration[188;191], cataract[184;187-189], primary open angle glaucoma[188], diabetic retinopathy[188], corneal transplantion[192] and central retinal vein occlusion[193].

2.2.6.8 Measuring HRQOL in paediatric ophthalmology

Despite the shortcomings of most HRQOL instruments and the lack of a specific instrument for paediatric ophthalmic patients it is important to try to evaluate the QOL of these children, as visual impairment affects all aspects of their lives. Most children with eye conditions have non-fatal chronic diseases requiring multiple hospital visits. The changing management of these children can be disruptive and include aggressive amblyopia treatment protocols, multiple operations with lack of certainty about the future compromising the quality of life for these children and their families. Importantly, as unilateral cataract is a non-blinding condition, it is essential that the management of these children is not doing more harm than good, overall.

The quality of life of these children would be expected to be reasonable as the disease is painless and although chronic has no fatal outcomes. Its association with other medical disorders may also complicate measuring the quality of life for children with congenital cataract, making it difficult to determine per se what the effect of the cataract on the lives of these children truly is.

At present there are no paediatric vision related quality of life instruments available. Furthermore there have been no previous studies investigating the HRQOL of children with any ophthalmic disorder on which the present study could be based. Thus for the present study, the PedsQL4.0™, a generic HRQOL questionnaire was used (Appendix 1). The PedsQL4.0™ comprises an 8 item physical health summary score (physical functioning) and a 15 item psychosocial health summary score (school, social and emotional functioning). A total score is ascertained from these. There are separate instructions for children aged 2-4, 5-7, 8-12 and 13-18 and children are asked about events over the past month.

Although the PedsQL4.0™, a generic questionnaire, lacks vision specific questions, it contains domains in which vision is important such as education. Other questions such as pain, are arguably less applicable. Importantly, the PedsQL4.0™ has parallel parent and child forms, is sensitive to cognitive development and has excellent internal consistency scores and clinical validity[194]. The PedsQL4.0™ requires minimum training or expertise to administer, is simple and fast to complete[194] and is applicable to children within the age range of this study. It is also free to use.

The PedsQL4.0™ items are rated on a 5 point Likert scale from 0 (never a problem) to 4 (almost always a problem). Scores are then transformed to a score ranging from 0-100 with higher scores representing a better QOL[195]. The original American version is currently in use although an anglicised version is being

developed, and the differences in language are anticipated to be minimal (Penny Upton, Research Associate, Department of Psychology, University of Sheffield: personal correspondence).

2.2.7 Summary of literature review

The literature review has highlighted several areas which require further research.

- No population based studies reporting the visual acuity of children with congenital cataract and investigation of predictors of visual acuity and postoperative complications.
- Very few studies exploring the causes of difficulties of amblyopia treatment .
- The quality of life of children with ophthalmic disorders has not been previously investigated. Furthermore the development of a paediatric vision related quality of life instrument is needed.
- No studies investigating the pathogenesis and aetiology of postoperative open angle glaucoma, and its risk factors.
- Further studies are required to investigate the longterm effectiveness and safety of IOL implantation especially in children operated at <2 years of age. Issues requiring further investigation include:
    - Ideal refraction at implantation, power and type of IOL
    - Effectiveness compared to traditional aphakia and CL correction
- Evaluation of the red reflex examination and implementation of training for screening paediatricians and its effectiveness
- Very few randomised control trials of congenital cataract

2.3 The British Congenital Cataract Study and the British Congenital Cataract Interest Group

The children comprising the cohort for this study were first identified and studied in 1995-1996 (Rahi J, PhD thesis 1998, University of London). The British Congenital Cataract Interest Group (BCCIG), comprising those ophthalmologists in the United Kingdom responsible for the management of young children with cataract was established in May 1995 (refer to *9.8 Appendix 8 Members of the BCCIG*). The group was established in order to undertake a national study of the incidence, mode of detection, causes and initial management of children newly diagnosed with congenital and infantile cataract in the UK (British Congenital/Infantile Cataract Study, BCCS). All children newly diagnosed with congenital/infantile cataract in the UK during the 12 months from October 1995 were identified using the national paediatric active surveillance scheme run by the British Paediatric Surveillance Unit and independently, but simultaneously, a disorder-specific active surveillance scheme in ophthalmology established through the BCCIG. The BCCS study cohort comprises 248 children and is considered to be nationally representative as capture-recapture analysis suggested that 92% of eligible infants were ascertained[28]. Detailed information, including initial management, was collected on all cases using standard forms for the year following diagnosis. A number of findings have been reported, including those regarding detection of children and the role of the child health screening and surveillance programme; age-specific and cumulative incidence and ascertainment of congenital cataract through the National Congenital Anomaly System; and underlying or associated causes of cataract[47;196-199].

2.4 Analysis of Ophthalmic Data

2.4.1 One Vs Two Eyes Analysis

The analysis of data in ophthalmology usually involves information on both eyes, as frequently disease affects both eyes. A disease may affect for example, the right eye in one individual and the left in another or a subject may refuse to have both eyes tested. Therefore, information may be available in one eye or both eyes and in either the right or left eyes of different people.

Some outcome measures are dependent on information from both eyes, and the analysis is then at the level of the person, such as the measure of stereopsis. Similarly if research is investigating treatment such as concordance with occlusion, then analysis at the level of the individual is appropriate.

Other measures such as visual acuity or intraocular pressure are made at the level of the eye, but in such variables there is usually a high degree of correlation between the two eyes of the same individual as well as in repeated measures in the same eye over time. One study simulating an exercise based on a comparison of introcular pressure found that failure to take into account between eye correlation led to an increase in the rate of false positives results from 5% to 20%[200].

The problem with correlation between eyes may be tackled by simply analysing only one eye of an individual. This however loses data and can introduce bias.

Similarly if an average of both eyes is used, it will lose data especially if correlation is low. The averaging of data may be acceptable on a person basis, but on an individual eye basis such as the impact of a type of surgery on visual acuity, it will again introduce bias.

Associations between eyes can be accounted for by clustering within the child and multi-level modeling approaches have been advocated as measures to address correlation between eyes[201].

2.4.2 Developmental changes in childhood

In the analysis of outcomes in children, normal developmental changes must be taken into account. For example, the age of the child will affect the best visual acuity obtainable as the development of the visual system at different ages will be variable (refer to *2.1.2 Visual development, page 21)*. Furthermore as the child matures, becomes literate and familiar with testing protocols the visual acuity will improve[201]. Analysis of data therefore needs to take into account the age of the child at assessment.

3 SUMMARY OF AIMS OF THIS STUDY ON THE OUTCOMES OF CONGENITAL CATARACT

Congenital cataract is a rare disorder but is a priority of Vision 2020, the international programme for the elimination of avoidable blindness, reflecting its potential for treatment and the consequences for the child and family if left untreated. As the aetiology of congenital cataract is unknown in the majority of cases with only a minority being preventable, informing secondary and tertiary approaches to prevent visual impairment due to congenital cataract is currently essential.

The British Congenital Cataract Study (BCCS) cohort comprises a nationally representative group of children who at the time of the present study, have all been under active management for at least 6 years after diagnosis. At this stage, all the most important outcomes of the whole range of prevailing management practices of children with congenital cataract may be accurately investigated. In addition, information about relevant risk factors, such as age at detection and at surgery, cataract morphology and presence of other ocular abnormalities were recorded prior to the present study, reducing the potential for bias and allowing a longitudinal approach to the analysis of the data.

Deficiencies in studies informing current management to date have been identified (refer to section *2.2.7 Summary of literature review, page 59*). This study aims to report the visual acuity of children with congenital cataract and to identify and quantify factors which predict visual acuity. Postoperative

complications, with notable emphasis on postoperative open angle glaucoma will also be reported and predictive factors will be identified and quantified..

The quality of life of children with congenital cataract has not previously been reported. The present study aims to investigate the feasibility of assessing the health related quality of life of children with congenital cataract and to compare their HRQOL with other childhood diseases.

Data are also limited on the impact of amblyopia treatment on the psychological wellbeing of the child with congenital cataract and their families. This study aims to explore the psychosocial impact of amblyopia treatment on the children and their families and investigate if psychosocial factors influence concordance with amblyopia treatment.

Therefore the research described in this thesis was undertaken to provide information on functional clinical outcomes, quality of life and the impact of amblyopia treatment in order to inform better current clinical practice and aid the evaluation of future and existing regimes. It will also form the basis of further research into under investigated outcomes such as vision related quality of life, as well as emerging therapeutic strategies.

4 METHODS

In this chapter, the identification and follow up of a nationally representative group of children diagnosed with congenital cataract in 1995/6 is described. The approaches used to collect outcome data including development and use of data collection instruments and application of a generic quality of life instrument are discussed. Statistical analyses undertaken are described together with issues relevant to analysis of paired (two eyes) ophthalmic data.

4.1 Study of national outcomes of congenital cataract

4.1.1 Identification of the children and their managing consultants

The managing consultant of each child at the last follow up visit from the original study (*refer to section 2.3 The British Congenital Cataract Study and the British Congenital Cataract Interest Group, page 60*) was sent a letter outlining the purposes of the present project and requesting their participation. The proposed method of collecting data by questionnaire completed by the managing consultant or a senior member of their team was introduced. A list of the child(ren) thought to be under the care of each consultant was sent to them for verification. Once a consultant had verified the child was or had been under their care and indicated that they would like to participate in the study, a questionnaire was sent for completion on each child. An offer of a visit to the unit by the principal investigator to complete the questionnaires on behalf of the consultant was made to all consultants with more than 5 children in the study. The consultants were also invited to suggest other ways in which the principal investigator could aid completion of the questionnaires.

The management status of the child was asked i.e. whether the child was still under the care of that consultant, had been discharged, lost to follow up or referred to another colleague. The consultant was asked to provide information on the new managing consultant if the child had subsequently been referred. If a child was no longer in the care of a consultant, the consultant was asked for

information from the most recent assessment up until the point they were discharged, referred or lost to follow up.

For any child under the care of a consultant who was not a member of the BCCIG, papers reporting findings from the original study were sent together with a letter inviting the consultant to join BCCIG and participate in the present study.

4.1.2 Defining outcomes and the design and development of questionnaires

A review of the literature was undertaken initially to identify outcomes of interest in relation to congenital cataract and factors which influence them. This was used as the basis for eliciting the opinions of consultant ophthalmologists initially at Great Ormond Street Hospital and then subsequently more broadly with members of the British Congenital Cataract Interest Group.

Following this, data collection forms were developed in consultation with epidemiologists with expertise in questionnaire design. These were circulated amongst BCCIG members prior to formal piloting. They were asked to comment on the questionnaire and suggest necessary amendments or additions. A prepaid envelope was provided for their reply. Only minor amendments were suggested which were incorporated. Finally, the questionnaires were formally piloted by the principal investigator and minor amendments to layout were made before general distribution to members of the BCCIG.

The forms were designed to encompass all important outcomes and to be applicable to all children in the study population. Sections of the questionnaire would therefore be left blank if not applicable to the child and could therefore be completed fairly quickly in the majority of children. A combination of open ended and closed questions were used and wherever possible, forced choice tick boxes were provided to ease completion. The provision of a pre-coded list of responses served to further clarify the question being asked.

The questionnaire was in 2 parts (Appendix 2):

Part 1: to be completed for all children

Verification on data already collected in the 1995/6 study.

Information on the current management status of the child, the findings at the last clinical examination and the details about investigations and amblyopia treatment were collected. There were also questions on the education and certification status of the child.

Part 2: to be completed only if applicable to the child

Questions in this section focused on surgery, if the child had undergone a surgical procedure. Details about the cataract surgery, intraocular lens implantation, and complications of cataract surgery including posterior capsular opacification and secondary membrane formation, postoperative uveitis, retinal detachment, glaucoma, endophthalmitis and strabismus were requested. Information on any other additional medical, laser or surgical procedures (including strabismus) was requested. There was also a section available to complete if there had been any other complications or surgical interventions not covered elsewhere in the questionnaire.

4.1.3 Data collection

Data was collected in an 8 month period from 1st September 2003 to the 1st May 2004. Prepaid envelopes were provided for return of completed questionnaires. On receipt of a questionnaire, the principal investigator sent a letter acknowledging receipt and thanking the managing team for their help in the study.

To encourage response, a high level of communication was maintained with all ophthalmologists who agreed to participate. As described earlier, at an early stage members of the BCCIG were consulted on the proposed methods of study including data collection to reduce their workload and ease completion of the questionnaire.

Reminder letters were sent after 8 weeks, followed by a phone call if there was still no response. Further questionnaires were sent as necessary. If the consultant was finding the completion of the questionnaires difficult, an offer was made by the principal investigator to visit the unit to complete the questionnaires on their behalf. Alternatively, ophthalmologists were offered the opportunity to send anonymised photocopies of relevant sections of the notes from which the principal investigator could complete the questionnaires.

Regular feedback through correspondence was maintained with members of BCCIG throughout the project and an end of project report was also sent.

4.1.4 Verification of Data

The completed questionnaires were scrutinised by the principal investigator for consistency and validity of the reported information. Where there was missing data or inconsistency, the respondent was contacted to verify information where possible.

4.1.5 Data protection and confidentiality

Although full identifiers (name, date of birth, address) had been available for the original study, in accordance with subsequent new guidance on data confidentiality, the children were only identifiable by a combination of their date of birth and the first three letters of their surname. Questionnaires were kept in a secure locked filing cabinet. The unique identifying number assigned to each child on the database of the original study was retained as described below.

4.1.5.1 Ethics approval

This study was approved by the Great Ormond Street Hospital/Institute of Child Health local research ethics committee. A further request was made and ethics approval granted allowing examination of the children for validation purposes.

4.1.6 Database management

Data from the original database was used in this study. Existing information included gender, date of birth, laterality, aetiology of the cataract, initial visual acuity, type of cataract surgery and early complications. Dates of events such as detection date and date of surgery were also available allowing possible longitudinal analysis of the data. The original database was a Microsoft Access (Microsoft Corp USA, 2003) database and the original numerical codes to uniquely identify the children were maintained. This allowed easy transfer and comparison of the data from the original to the present study.

Following consultation with statisticians and database experts, a new relational database was constructed in Microsoft Access. Numerical and categorical codes were assigned to the variables. Free text was also entered without coding in variables which were largely descriptive or had a large number of possibilities. The database was constructed to resemble the actual questionnaires to facilitate ease of entry and pull down menus were used in questions with closed ended answers. There were also internal checks to prevent entry of variables outside the acceptable range. Separate tables were constructed on clinical examination findings and other tables had information on surgical procedures and complications. The tables were then all linked by the original unique identifying numbers.

As each questionnaire was received, the data was entered into the outcomes database. A separate database was kept with the details of the consultant and how the consultant was progressing in completing the questionnaires.

4.1.7 Data entry

Numerical codes were assigned to items on the questionnaire. Free text responses and descriptive data were entered without coding, for example descriptions on other medical disorders. The databases were backed up to compact disc or Zip discs each day after data entry. A full back up of the computer was carried out each week.

4.1.8 Validation of Data

Where possible, for validation purposes, units with a large number of children (>4) were approached through the initial introductory letter regarding the feasibility of visiting the units and examining the children to allow independent assessment of outcomes. To minimise disruption, the principal investigator visited the units and examined the children in the context of a routine outpatient appointment wherever possible. However, some ophthalmologists also organised clinics specifically for this purpose. The data collected by clinical assessment were cross-referenced by reference to the hospital notes.

The examination of the children included visual acuity, stereopsis and strabismus assessment as well as a slit lamp examination. Parents were also questioned on issues such as education and social help for their child.

4.1.9 Verification of data entry

In alphabetical order, every fifth child's questionnaire was reviewed and the entry in the database was checked. The database had multiple entries for the same data in different sections so it was possible to cross check that data had been entered correctly. For example, if the child had an intraocular lens implanted at the time of surgery (entered in section on cataract surgery) there should also have been data on the details of the lens (entered in intraocular lens section) in the database. Of 50 questionnaires reviewed, 2 had minor mistakes only.

4.2 Health related quality of life and amblyopia treatment study

4.2.1 Identification of the children

All parents and their children were eligible for inclusion in this additional study. To maximize representation of the management of children with congenital cataract nationally, certain units which managed the largest numbers of children were targeted for the HRQOL and amblyopia studies. These children were to be also clinically examined as part of data validation (*refer to section 4.1.8 Validation of Data, page 73*). 42 pairs of children and their parents from 6 ophthalmic units nationwide (*refer to section 5.1.1 Distribution of children amongst managing consultant ophthalmologists, page 94*) were invited at their routine ophthalmology clinic appointment or the special clinics organised for the study to participate in the quality of life and amblyopia treatment study.

4.2.2 The design and development of questionnaires

4.2.2.1 Paediatric quality of life questionnaire (PedsQL 4.0^{TM})

Permission was sought and granted by the copyright authors for the use of PedsQL 4.0 ^{TM} (J. Varni, USA 2002). The PedsQL 4.0^{TM} questionnaires were reproduced according to standard design and were colour coded (white for the parental version and yellow for the child version) to aid recognition. The bright yellow colour was also thought to be more appealing to the children. The smallest font size was 14 and each section was numbered and titled to aid completion (Appendix 1).

4.2.2.2 Amblyopia treatment study questionnaire design

After consultation with consultant ophthalmologists, the head orthoptist and the head optometrist at Great Ormond Street Hospital as well as psychologists and questionnaire experts, a questionnaire was designed to study the impact of wearing contact lenses, glasses and occlusion on the life of the child and their relationships with family and friends (Appendix 3). The questionnaire was very loosely based on the questionnaire used in the Amblyopia Treatment Trial undertaken in Newcastle by Mr M Clarke, consultant ophthalmologist. It was designed to be completed by the parents and was colour coded light blue to aid recognition. Initial questions asked about the age at the commencement of treatment, open ended questions on preference of glasses to contact lenses (or vice versa) and any behavioural changes noted as a consequence of amblyopia treatment. The final questions used a 5-point Likert scale and were asked to ascertain the feelings on the experience of the parent and the child during amblyopia treatment, their relationships with their friends and family and the impact that treatment had on their abilities at school. The smallest font size was 14 and each section was numbered and titled to aid completion.

4.2.2.3 Piloting of questionnaires

The questionnaire was initially piloted amongst 24 parent-child pairs at Great Ormond Street Hospital. Overall the response was positive and parents and children were willing and able to participate. In addition, visually impaired parents and children were specifically asked if they had any problems in reading the questionnaire. Only one parent reported a problem and this was overcome by reading it more slowly.

4.2.3 Data collection

To enhance participation, the study was fully discussed with the parents who were given ample opportunity to ask questions or withdraw. The parents consented to participation to the clinical examination and questionnaire at this clinic visit unless they had been approached previously by letter. Questionnaires were completed in clinic if space and time permitted. Parents and children completed the questionnaires independently (in the same room, but at opposite ends) and the investigator was present to answer any queries. If the questionnaire was not completed in clinic, but consent had been given, the questionnaire was posted to the parent and child for completion at home. For the postal questionnaires, a prepaid envelope was provided for their return.

Parents and children who were completing the questionnaires at home were encouraged to respond. Reminder letters were sent out after 6 weeks and then a

follow up phone call if there was still no response. Further questionnaires if necessary were also sent out. If the family was finding the completion of the questionnaires difficult, an offer was made to visit the home to help complete the questionnaires or they would bring in the questionnaires at the next routine clinic appointment.

When a questionnaire was received, a letter was sent acknowledging receipt and thanking the parent and child for their help in the study.

4.2.4 Verification of Data

The completed questionnaires were scrutinised for consistency and validity of the reported information. Where there was missing data or inconsistency, this was classified as 'missing'.

4.2.5 Ethics approval

A specific request was made and approval granted by the Great Ormond Street Hospital/Institute of Child Health local research ethics committee for recruitment of the children to assess quality of life and the impact of amblyopia treatment.

4.2.6 Database management

Following consultation with statisticians and database experts, two new relational databases were constructed in Microsoft Access for the data from the PedsQL 4.0™ and amblyopia treatment study. Numerical and categorical codes were assigned to the variables. Internal checks in the database prevented entry of variables outside the acceptable ranges. The databases were linked to the original cohort database by the unique identifying number assigned to each child.

As each questionnaire was received, the data were entered into the PedsQL4.0™ and amblyopia treatment study databases. A separate database was kept with the contact details of the family and how the family was progressing in completing the questionnaires.

4.2.7 Data entry

Numerical codes were assigned to items on the questionnaire. Free text responses and descriptive data were entered without coding, for example situations in which the child preferred wearing contact lenses. The databases were backed up to compact disc or Zip discs each day after data entry. A full back up of the computer was carried out each week.

4.2.8 Verification of data entry

All entries in the database were reviewed a second time to check for errors. 3 had minor mistakes, which were corrected.

4.3 Statistical Analysis

4.3.1 Preparation of data for analysis

Data from the Microsoft Access database (Microsoft Corp, USA 2002) was formatted to be able to be readily transferred into SPSSv11 (SPSS Inc, USA 2003) for analysis. This involved recoding some variables and limiting the size of others as described below.

Two new databases were created in SPSSv11. One database had a line assigned per child and in the other database each line represented an eye of a child (ie a child was assigned two lines: one for the right eye and one for the left eye). This allowed analysis of the data both by person and by eye.

4.3.2 Generation of new variables for analysis

4.3.2.1 Time since date of presentation, date of surgery and date of occurrence of complications

With the known dates of significant events such as the date of presentation, date of surgery and date of occurrence of complications, it was possible to determine time intervals and generate these as further variables for analysis.

4.3.2.2 Visual acuity (VA)

Due to the variation in testing methods in the different units and the large number of possible visual acuities, the recorded visual acuity measures were refined to allow analysis of the data as described below. The majority of children had been recorded using Snellen visual acuity.

A standard Snellen chart contains a 6/60 line with 1 letter, 6/36 line with 2 letters, 6/24 line with 3 letters, 6/18 line with 4 letters, 6/12 line with 5 letters, 6/9 line with 6 letters, 6/6 line with 7 letters, 6/5 line with 8 letters, 6/4 line with 9 letters. The VA was converted into whole lines rather than including part lines. This rounding up/down took into account the number of characters per line. If the VA was recorded as having read over half of the letters in the next successive line, then the VA was recorded as the next line up (e.g.6/12+4 was recorded as 6/9). If the VA was recorded as having read less than half of the letters in the next successive line then the VA was recorded as that line (e.g.6/12+2 was recorded as 6/12).

If the VA had been recorded as logMAR,which would have been more precise, these were nevertheless converted into Snellen acuity because the majority of children's visual acuity was recorded as Snellen. This permitted visual acuity outcomes to be treated in an ordinal fashion, in particular the worst acuities (and therefore the ones of most interest) which were not on a numerical scale (for example hand movements, perception of light, no perception of light).

4.3.2.3 Surgical procedure

The surgical procedure used by the consultants varied and included descriptions of machines and equipment. Therefore, in consultation with Professor David Taylor at Great Ormond Street Hospital, all surgical techniques reported were recoded into one of three mutually exclusive groups. This ensured that clinical issues surrounding surgical management of children with congenital cataract would be addressed whilst permitting meaningful analysis. Firstly, lensectomy/vitrectomy: removal of all lens material and capsule and some vitreous with the use of a vitrectomy machine. Secondly, lens aspiration alone: any lens aspiration technique, phacoemulsification or extracapsular cataract extraction with an anterior capsulorrhexis, +/- intraocular lens implantation. Thirdly, lens aspiration and posterior capsulorhexis: any lens aspiration technique, with an anterior capsulorrhexis, posterior capsulorrhexis and vitrectomy.

4.3.2.4 Postoperative complications

Whilst retaining information on actual complications, postoperative complications were also recoded into a new category of either sight threatening and non sight threatening categories. This was due to their diversity and rarity of their occurrence and allowed further analysis.

4.3.2.5 Additional non-ophthalmic disorders

Although the impact of any additional non-ophthalmic disorder varied, each was dichotomised into presence or absence. This allowed further analysis of descriptive data.

4.3.2.6 Severity of cataract

The morphology of the cataract was varied. The cataract was graded as severe if recorded as such in the clinical notes or implied by early cataract surgery. This was to allow further analysis of the data.

4.3.2.7 Aetiology of cataract

Remaining consistent with the original study, the children were assigned to one of three clinically relevant and mutually exclusive categories by the aetiology of their cataract. Firstly, isolated cataract (isolated), secondly, cataract specifically associated with ipsilateral ocular disorder without any systemic disease (ocular). Thirdly, cataract specifically associated with specific systemic disorder with ocular disease(ocular systemic) and finally, cataract associated with specific systemic disorder with no ocular disease (systemic).

4.3.3 Analysis of data

Descriptive analyses of outcomes of interest and of possible associated factors were undertaken initially. Analysis of the amblyopia treatment study was descriptive only.

4.3.3.1 Outcomes of interest

4.3.3.1.1 Visual Acuity

Children with unilateral cataract were analysed separately from children with bilateral cataract. The eyes with cataract of children with unilateral cataract were assessed only, as these represented the outcomes of interest (*refer to section 2.2.2 Management of congenital cataract by the laterality of the cataract, page 34).*

A priori, correlation between potential predictors (time since detection, age at detection, time since cataract surgery, age at cataract surgery, type of refractive correction, type of cataract surgery, primary intraocular lens implantation, concordance with occlusion regime, aetiology of cataract, presence of non-ophthalmic disorders, severity of cataract at presentation, sight threatening postoperative complications and gender) was analysed using parametric and nonparametric tests (chi- squared, Mann Whitney U, Kruskal- Wallis and Spearman's correlation coefficient). Next, using ordinal regression, univariate association between predictors and visual acuity were estimated. Subsequently,

combinations of factors were used (backwards, forwards and enter strategies) to yield the simplest multivariate model that best predicts visual acuity. To identify reverse confounding, the final model was reassessed with those variables which had originally not been significant univariately but which were biologically plausible.

The correlation between eyes of paired eye data in visual acuity analyses was adjusted by clustering within child using STATA(Stata corp USA, 2003) software.

4.3.3.1.2 Glaucoma

Descriptive analysis of all types of glaucoma was undertaken initially.
Postoperative open angle glaucoma was analysed separately as it is emerging as potentially the most sight threatening post operative complication of surgery for congenital cataract.
Postoperative open angle glaucoma for the purposes of this study was defined as:

Any child who underwent continual sustained treatment for glaucoma with drops and /or laser and/or surgery and having been clinically diagnosed by the managing consultant as having postoperative open angle glaucoma

A priori, correlation between potential predictors (age at detection, age at cataract surgery, cataract surgery procedure, vitrectomy at primary procedure,

primary intraocular lens implantation, significant postoperative uveitis, microphthalmia, severity of cataract at presentation, gender and laterality) was analysed using parametric and nonparametric tests (chi- squared, Mann Whitney U, Kruskal- Wallis and Spearman's correlation coefficient).

Next, using Cox regression, univariate association between predictors and postoperative open angle glaucoma were estimated. Subsequently, combinations of factors were used (backwards, forwards and enter strategies) to yield the simplest multivariate model that best predicts postoperative open angle glaucoma. To identify reverse confounding, the final model was reassessed with those variables which had originally not been significant univariately but which were biologically plausible.

The correlation between eyes of paired eye data in glaucoma analyses was adjusted by clustering within child using STATA(Stata corp USA, 2003) software.

4.3.3.1.3 Pediatric Quality of Life

The distribution of the physical, psychosocial and total summary scale scores was assessed to determine whether parametric tests could be applied. Internal consistency was measured using the Alpha Cronbach Coefficient scores[202] and the Bland Altman measure of agreement[203] was used to assess parent-child agreement. Scores obtained in this study were compared to other published studies in the literature using the PedsQL 4.0 ^{TM} to measure the HRQOL of children with other disorders.

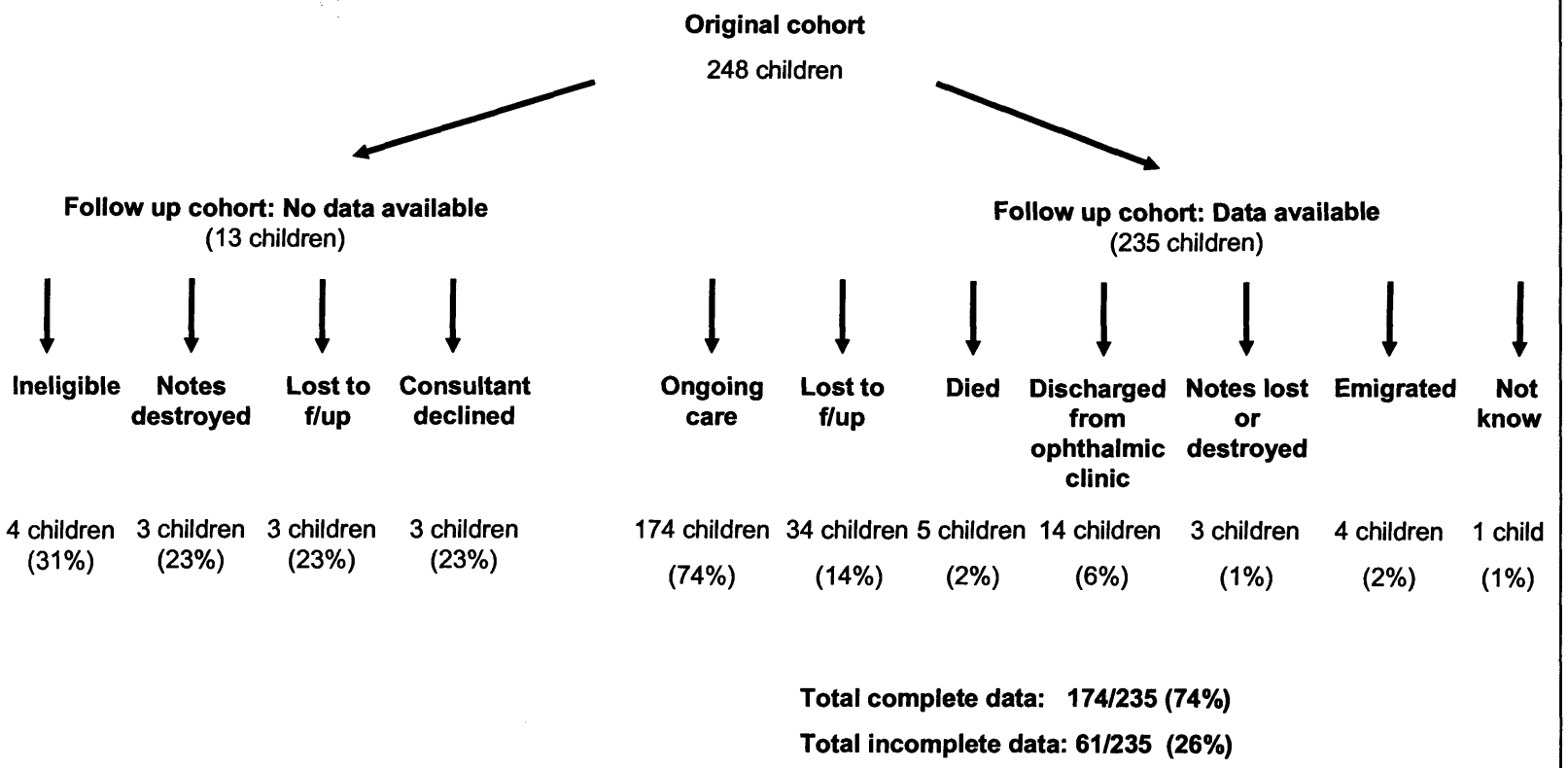
A priori, the influence on QOL scores of age, sex, laterality of cataract, presence of other medical conditions, surgery, strabismus, significant postoperative complications, glaucoma, posterior capsular opacity or amblyopia treatment was examined using analysis of variance and of best visual acuity (regression analysis) where best visual acuity was defined as best visual acuity either monocularly or binocularly

5. RESULTS

5.1 Completeness of follow up and data collection

Questionnaires were completed for 235/244 (96%) of eligible children in the study. In total, 298 questionnaires were completed as some children had data from more than 1 source (Figure 1).

Figure 1: Completeness of data and follow up of children in this study



A reply was received from all consultants (102) who were approached to participate in the study with only one declining to participate. Of these, the principal investigator completed 211(71%) questionnaires on 178 children. This included visiting 17 units recording data from patients cared for by 24 consultants (Table 3) and 9 questionnaires were completed from photocopied notes (Table 4). In 6 of the 17 units visited, the principal investigator undertook clinical assessments of the children for data validation (Table 5). 87(29%) questionnaires were completed by the managing consultant or a senior member of their team.

Table 3: The units visited for completion of questionnaires by principal investigator

Hospital Visited	Number of questionnaires N=192 (Number of children=169)
Birmingham Children's Hospital	12
Bradford Infirmary	4
Bristol Eye Hospital	13
Leighton Hospital, Crewe	2
Kent County Ophthalmic and Aural Hospital, Kent	1
Charing Cross Hospital, London	1
Great Ormond Street Hospital for Sick Children, London	78
King's College Hospital, London	3
Leicester Royal Infirmary	8
Moorfield's Eye Hospital, London	22
The General Infirmary, Leeds	3
Alder Hey Children's Hospital, Liverpool	10
Manchester Eye Hospital	19
Milton Keynes Hospital	4
Queen's Medical Centre, Nottingham	4
The Royal Victoria Infirmary, Newcastle	7
Southampton Hospital	10

Table 4: The units who sent photocopies of the notes for the questionnaires to be completed by the principal investigator

Hospital	Number of questionnaires N=9 (Number of children=9)
Royal Aberdeen Children's Hospital	5
Manchester Eye Hospital	2
The General Infirmary, Leeds	2

Table 5: The units visited by the principal investigator for data validation

Hospital Visited	Number of children examined N=42
Birmingham Children's Hospital	4
Bristol Eye Hospital	6
Leicester Royal Infirmary	3
Great Ormond Street Hospital for Sick Children, London	21
Moorfield's Eye Hospital, London	3
The Royal Victoria Infirmary, Newcastle	1
Southampton Eye Hospital	4

Thus, there was data available in 235/244 (96%) children eligible for this study, in 74% of whom had complete data. Therefore, the denominators for different data items will vary and are reported separately. Table 6 compares eligible children (N=235) included in the study with those not included (N=9). Gender and laterality distribution appears to be very similar, but only 2 categories of aetiology were represented in the excluded group making further comparison difficult. The management of the children not included (ie surgery or no surgery) is not known.

Table 6:Comparison of eligible children included and not included in this study

	Included in study (N=235)	**Not included in study (N=9)**
Gender	Boys 122 (52%) Girls 113 (48%)	Boys 5 (56%) Girls 4 (44%)
Laterality	Bilateral 158/235 (67%) Unilateral 77/235 (33%)	Bilateral 7/9 (78%) Unilateral 2/9 (22%)
Aetiology	Isolated 130/235 (55%) Systemic 31/235 (13%) Ocular 63/235 (27%) Ocular Systemic 11/235 (5%)	Isolated 6/9 (67%) Systemic 3/9 (33%)

5.1.1 Distribution of children amongst managing consultant ophthalmologists

The original cohort comprised 248 children who were managed by 64 consultant ophthalmologists nationwide. By the time of the present study, one ophthalmologist declined to be part of the study, 11 ophthalmologists had retired or died or no longer looked after the paediatric patients in their departments and 38 new ophthalmologist joined BCCIG, making a total of 90 consultants.

The majority of ophthalmologists 57/90 (63%) cared for one child (Table 7). Most of these were managing children referred back to their local hospital for long-term follow up either following surgery elsewhere or because they had stable mild to moderate cataract not requiring surgery. 48/248 (19%) of children were managed at Great Ormond Street Hospital at the time of their last follow up, whilst 17 other consultants managed between 4 and 19 children.

Table 7: The number of children cared for by each ophthalmologist

No. of cases per ophthalmologist (N=248)	No. of ophthalmologists (N=90)	Percentage of total number of cases (N=100)
1	57	23
2	13	10
3	3	4
4	5	8
5	5	10
7	1	3
8	1	3
9	1	4
10	2	8
19	1	8
48	1	19

5.2 Descriptive analysis of the study population

122/235 (52%) of the children in the present study were boys. 158/235 (67%) had bilateral cataract and 77/235 (33%) had unilateral cataracts (40 left sided, 37 right sided). Mean age at follow up of children with bilateral cataracts was 7.49 years (0 to 20 years) and those with unilateral cataracts was 6.64 years (0 to 12 years)

In 215/235 (91%) children the date of the last follow up visit was recorded and length of follow up could be determined. Time since the point of detection of the cataract ranged from 0 to 7.94 years (median=6.30 years).

Cataract morphology varied by laterality as shown in Table 8. In 4 bilateral cases morphology differed between the 2 eyes.

Table 8: Morphology of cataract (data available on 122/158(77%) bilateral cases and 62/77(81%) unilateral cases

Morphology	Laterality of cataract			Total
	Bilateral right	Bilateral left	Unilateral	
Total	19	19	10	48
Nuclear	31	31	7	69
Nuclear & post subcapsular	1	1	1	3
Nuclear and lamellar	2	2		4
Nuclear and cortical	2	1		3
Cortical	7	8	4	19
Posterior cortical		1	2	3
Anterior cortical			1	1
Lamellar	28	28	2	58
Lamellar & post lenticonus	1	1		2
Anterior Capsular	1	1		2
Anterior polar	6	6	6	18
Ant & post central plaques			1	1
Posterior polar	5	5	2	12
Post polar & post lenticonus	1	1		2
Posterior subcapsular	12	12	12	36
Posterior dumbbell	1	1		2
PHPV	1	1	1	3
Mild dot	3	3	1	7
Bluedot	1	1		2
Off visual axis morph NK			1	1

14/77 (18%) of children with unilateral cataracts and 13/158 (8%) children with bilateral cataracts had microphthalmia. Children with bilateral cataracts and microphthalmia had microphthalmia in both eyes.

Severity of the cataract at presentation was dichotomised into severe and non severe as recorded by the clinician or implied by immediate extraction of the cataract (Table 9).

Table 9: Severity of the cataract (by eye)

		Laterality of the cataract		Total
		Bilateral (N=316)	Unilateral (N=77)	
Severity of the cataract at presentation	Severe	217	15	232 (59%)
	Not severe	99	62	161(41%)
Total		158	77	393

Aetiology of the congenital cataract at time of follow up is shown in Table 10, using the classification described earlier (*refer to section 4.3.2.7 Aetiology of cataract, page 83*). Although the aetiology was different between the eyes of 2 bilateral cases, they were still put in the same category of this classification

Table 10: Classification of congenital cataracts (by child)

		Laterality of cataract		Total
		Bilateral N=158	Unilateral N=77	
Cause or associated factor for congenital cataracts	Isolated	98(62%)	32(42%)	130(55%)
	Ocular	23(15%)	40(52%)	63(27%)
	Ocular Systemic	7(4%)	4(5%)	11(5%)
	Systemic	30(19%)	1(1%)	31(13%)

In 8 children the aetiology was revised during follow up and further assessment as shown in Table 11.

Table 11: Revised underlying or associated factors with congenital cataract in 8 children

Original aetiology	Number of children (N=10)	Development/change in aetiology
Multiple cardiac and limb abnormalities	1	Autosomal recessive congenital cataracts, language and hearing disability
Smith Lemli Opitz 1	1	Misdiagnosed with SLO1, problems with global development
Autosomal dominant	1	Chromosome 10 balanced translocation
Idiopathic cataract	1	Autosomal recessive congenital cataract (brother diagnosed with cataract)
Idiopathic cataract	1	Lowe's syndrome
Idiopathic cataract	1	Marinesco-Sjogren's Syndrome
Idiopathic cataract	1	Rhizomelic chondrodysplasia punctata
Idiopathic cataract	1	Late report of possible maternal rubella exposure at 12/40 weeks

At follow up, 47/158 (30%) of children with bilateral cataracts and 12/77 (16%) of children had other non-ophthalmic medical disorders. Of these, nine children developed new non-ophthalmic medical disorders since initial diagnosis (Table 12).

Table 12: Development of other non-ophthalmic medical disorders since diagnosis

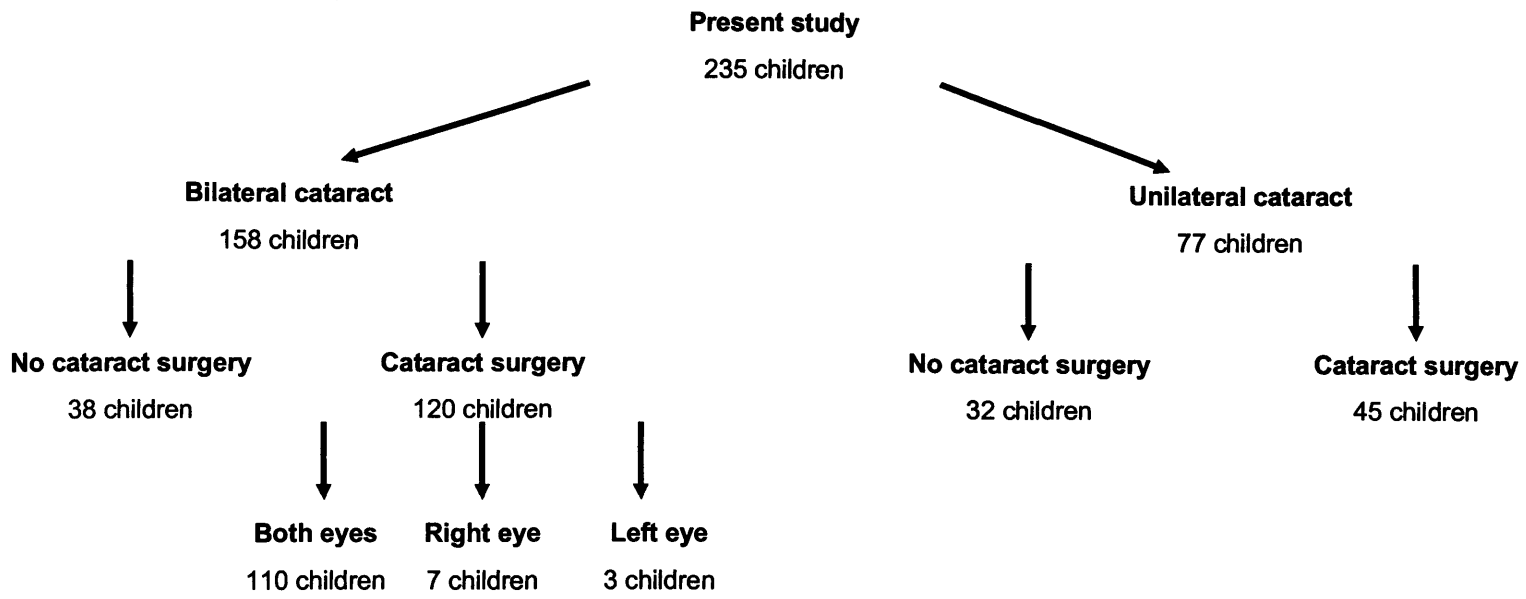
Aetiology of cataract	Number of children	Non-ophthalmic medical disorder
Idiopathic cataract	1	SLE and sickle cell anaemia
Idiopathic cataract	1	Sensorineural deafness
Idiopathic cataract	1	Partial deafness
Idiopathic cataract	1	Dislocated hips
Idiopathic cataract	1	Language disability and dental anomalies
Idiopathic cataract	1	Dyspraxia
Idiopathic cataract	1	Partial seizures
Idiopathic cataract	1	Dental anomalies
Autosomal recessive	2	Autism
Total	9	

5.2.1 Surgical management

5.2.1.1 Cataract surgery

Figure 2 shows the surgical and conservative management of the children in this study.

Figure 2: Surgical and conservative management of children with congenital cataract



Total number of eyes undergone cataract surgery = 230
Total number of eyes no cataract surgery = 76

Total number of eyes undergone cataract surgery = 45
Total number of eyes no cataract surgery = 109

Table 13 shows the types of cataract surgery performed. In bilateral cases the same procedure was performed on both eyes if both cataracts were extracted.

Table 13: Number of eyes undergoing different cataract surgery procedures (data available in 221/230(96%) eyes of bilateral cases and 45/45 eyes of unilateral cases)

		Laterality of cataract		Total
		Bilateral N=221	Unilateral N=45	
Type of cataract surgery	Lensectomy/vitrectomy (Cataract surgery age)	103 (47%) (5-2501 days median=63)	22 (49%) (15-1447 days median=57)	125
	Lens aspiration alone (Cataract surgery age)	72 (33%) (8-4425 days median=959)	14 (31%) 19-1744 days median=1040)	86
	Lens aspiration and vitrectomy (Cataract surgery age)	46 (21%) (27-2870 days median=799)	9 (20%) (43-2066 days median=862)	55

The age at cataract surgery by eye is shown grouped at three monthly intervals for children with bilateral cataracts in Figure 3 and unilateral cataracts in Figure 4.

Figure 3: Age at surgery by eye, of children with bilateral cataracts (N=215)

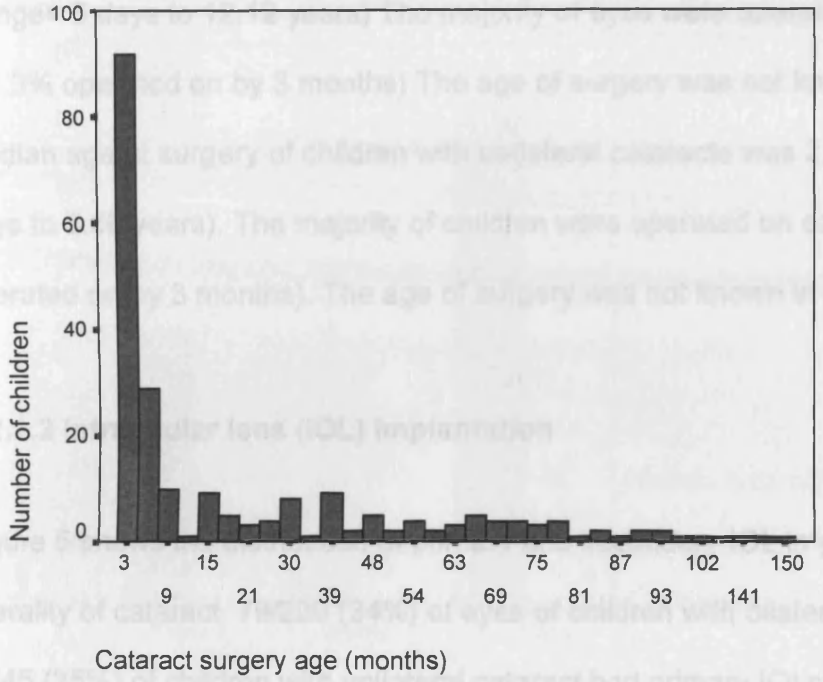
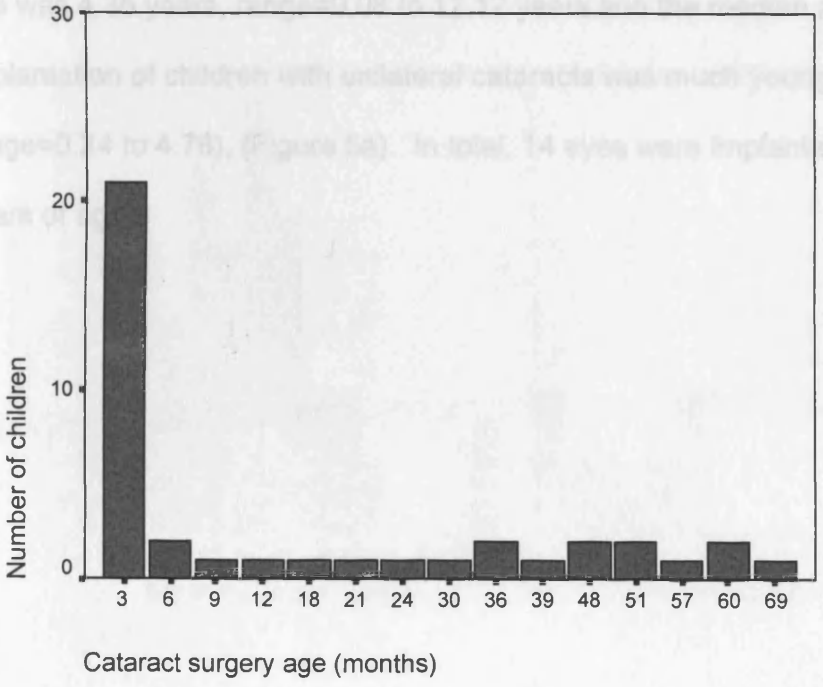


Figure 4: Age at surgery by eye of children with unilateral cataracts (N=40)



Median age at surgery of children with bilateral cataracts was 4.44 months (range= 5 days to 12.12 years) The majority of eyes were operated on early (41.3% operated on by 3 months) The age of surgery was not known for 7 eyes. Median age at surgery of children with unilateral cataracts was 2.8 months (15 days to 5.66 years). The majority of children were operated on early (52% operated on by 3 months). The age of surgery was not known in 5 children.

5.2.1.2 Intraocular lens (IOL) implantation

Figure 5 shows the distribution of primary and secondary IOL implantation by the laterality of cataract. 79/230 (34%) of eyes of children with bilateral cataracts, and 16/45 (35%) of children with unilateral cataract had primary IOLs implanted. The median age at surgery for IOL implantation of children with bilateral cataracts by eye was 4.35 years, range=0.08 to 12.12 years and the median age of implantation of children with unilateral cataracts was much younger (2.36 years, range=0.24 to 4.78), (Figure 5a). In total, 14 eyes were implanted at under 2 years of age.

Figure 5: Timing of IOL implantation by eye (N=Bilateral 230, Unilateral 77)

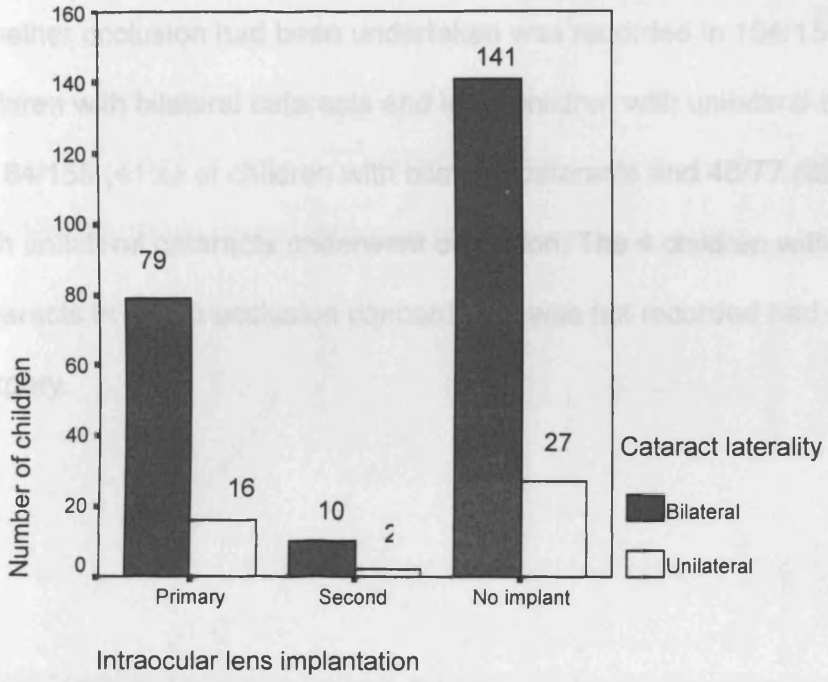
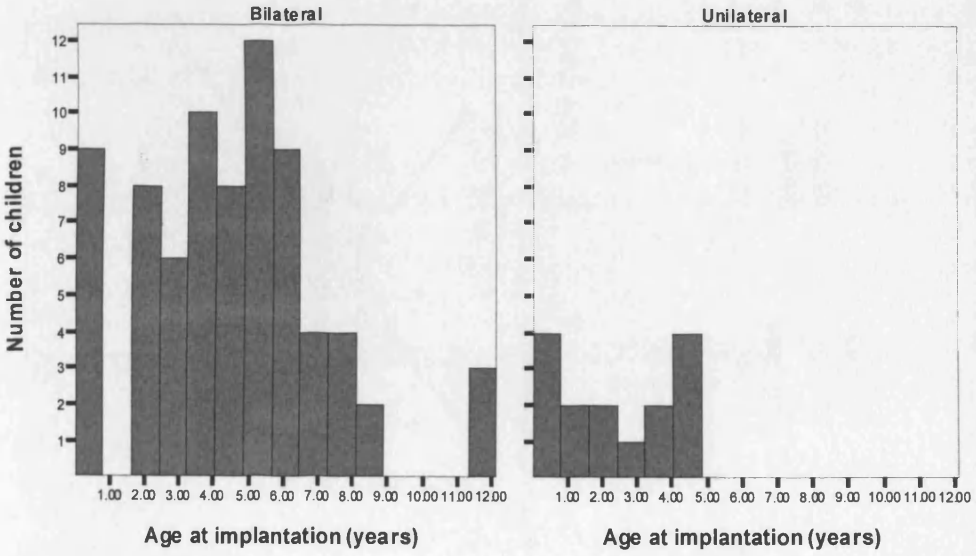


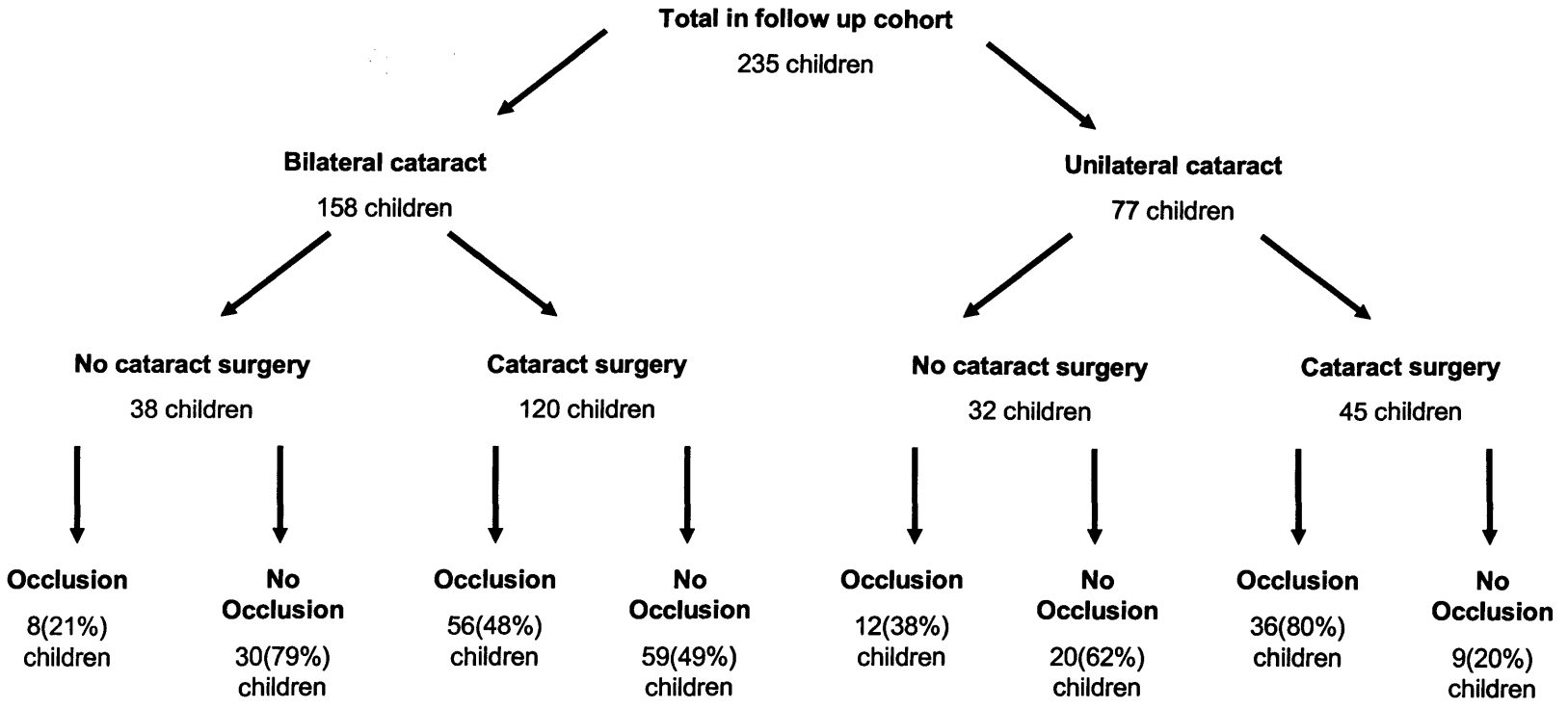
Figure 5a: Age of implantation of primary IOL by eye (N=90)



5.2.2 Non-surgical management

Whether occlusion had been undertaken was recorded in 154/158 (97%) of children with bilateral cataracts and in all children with unilateral cataracts (Figure 6). 64/158 (41%) of children with bilateral cataracts and 48/77 (62%) of children with unilateral cataracts underwent occlusion. The 4 children with bilateral cataracts in whom occlusion concordance was not recorded had undergone surgery.

Figure 6: Occlusion treatment of all children



NB: 4 children with bilateral cataracts not known if occluded

36/64(58%) children with bilateral cataracts (Table 14) and 27/48(56%) children with unilateral cataracts (Table 15) did not manage to achieve full concordance with their occlusion regime. The level of concordance was not known in 6 children with bilateral cataract.

Table 14: The percentage concordance with the occlusion regime that was achieved with children with bilateral cataracts undergoing occlusion (data available on 7/8(88%) children who did not have surgery and 51/56(86%) of children who did)

		Cataract surgery No N=7	Cataract surgery Yes N=51	Total 107
Percentage concordance with occlusion regime	<50	2(25%)	13(23%)	15(23%)
	50	1(13%)	5(9%)	6(9%)
	75	1(13%)	14(25%)	15(23%)
	100	3(38%)	19(34%)	22(34%)

Table 15: The percentage concordance with the occlusion regime that was achieved with children with unilateral cataracts (data were available on all children)

		Cataract surgery No N=12	Cataract surgery Yes N=36	Total
Percentage concordance with occlusion regime	<50	4(34%)	9(25%)	13(26%)
	50	1(8%)	6(24%)	7(15%)
	75	1(8%)	6(24%)	7(15%)
	100	6(50%)	15(42%)	21(44%)

53% of children with bilateral cataracts and 48% of children with unilateral cataracts prematurely terminated their occlusion regimes during treatment for the reasons shown in Table 16.

Table 16: Reasons for premature termination of occlusion treatment (data were not available for 3 bilateral and 2 unilateral cases)

Reasons for premature termination		Laterality of cataract		Total
		Bilateral N=61	Unilateral N=46	
Not terminated prematurely N=55		30(47%)	25(52%)	55(49%)
Terminated prematurely N=52	Parental and clinician decision	8(13%)	9(19%)	17(15%)
	Lost to follow up	2(3%)	1(2%)	3(3%)
	Clinician decision	15(23%)	10(21%)	25(22%)
	Glaucoma	2(3%)	1(2%)	3(3%)
	Parental decision	2(3%)		2(2%)
	Not known why abandoned	2(3%)		2(2%)

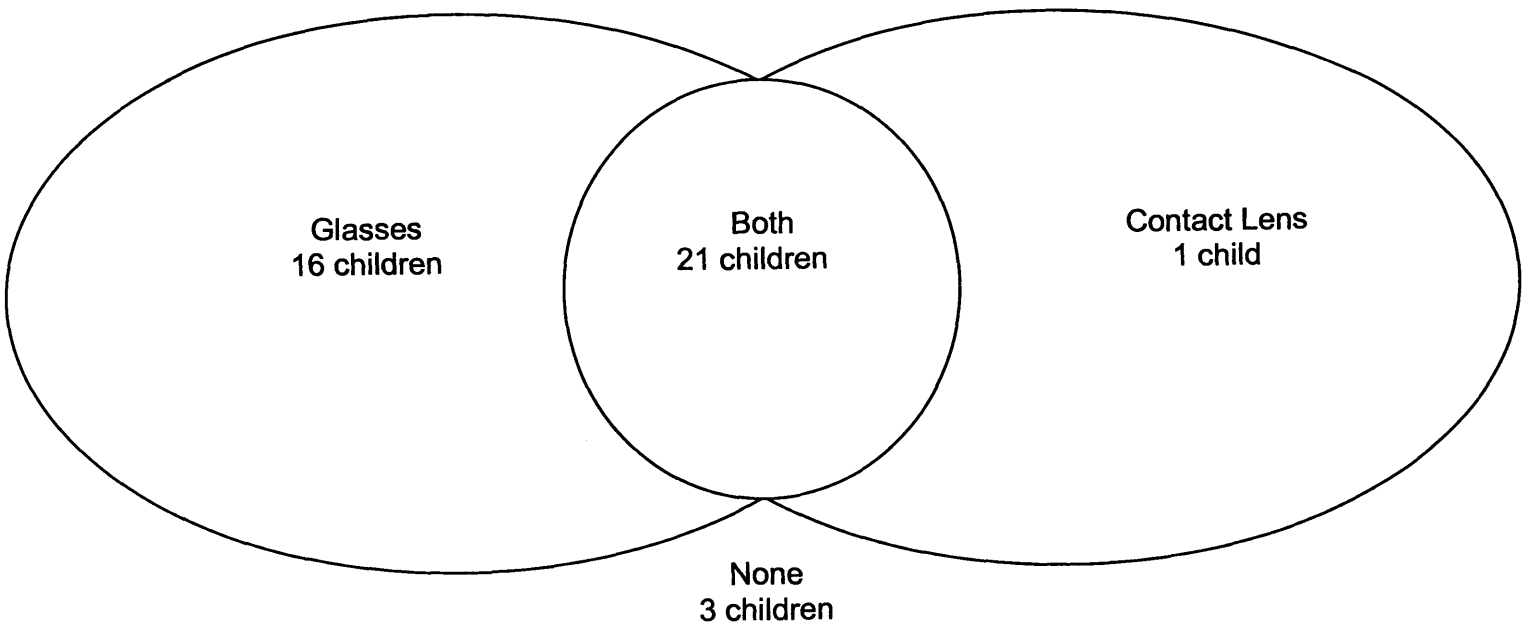
5.2.2.1 Parental perception of the effects of amblyopia treatment on their child

41/42 parents invited to participate completed questionnaires on amblyopia treatment. 6/41(19%) of the children had unilateral cataracts. The median age of participating children was 6.8 years (range 5.7-19.51), 24(59%) were girls and 13(32%) of all children had other medical conditions. The visual acuity scores (best obtained) of the children ranged from 6/4 to 6/72. (unilateral: 6/4-6/9, median=6/6; bilateral: 6/4-6/72, median=6/12. 34/41 (83% had undergone surgery).

The combinations of refractive correction worn by the 41 children are shown in Figure 7. Of the 22/41 children who were wearing or wore contact lenses, 14(64%) were girls. Median age of the child at time of questioning was 6.61 years(6.12 -12.02 years). Median age of starting to wear contact lenses (as recalled by parents) was 3.5 months(2 weeks to 3 years).

Of the 38/41 children who were wearing or wore glasses (reading and/or aphakic), 22(59%) were girls. Median age of the child at time of questioning was 6.72 years(5.63-12.19 years). Median age of starting to wear glasses (as recalled by parents) was 2 years(1 month to 3 years).

Figure 7: Venn diagram showing contact lens and glasses wear



5.2.2.2 Contact lens wearers

82% of the children wore the contact lenses as recommended by the clinicians. Most children (61%) reacted positively or very positively (scale point 4 or 5) to contact lens wear (Figure 8), despite 50% of parents finding the experience of their child wearing contact lenses difficult or very difficult (Figure 9). Furthermore, parental perception of CLs appeared to be good, as 71% of parents never or occasionally worried that the CLs were causing harm to their child (Figure 10).

Figure 8: Child's reaction to wearing CLs N=17 (5 not known)

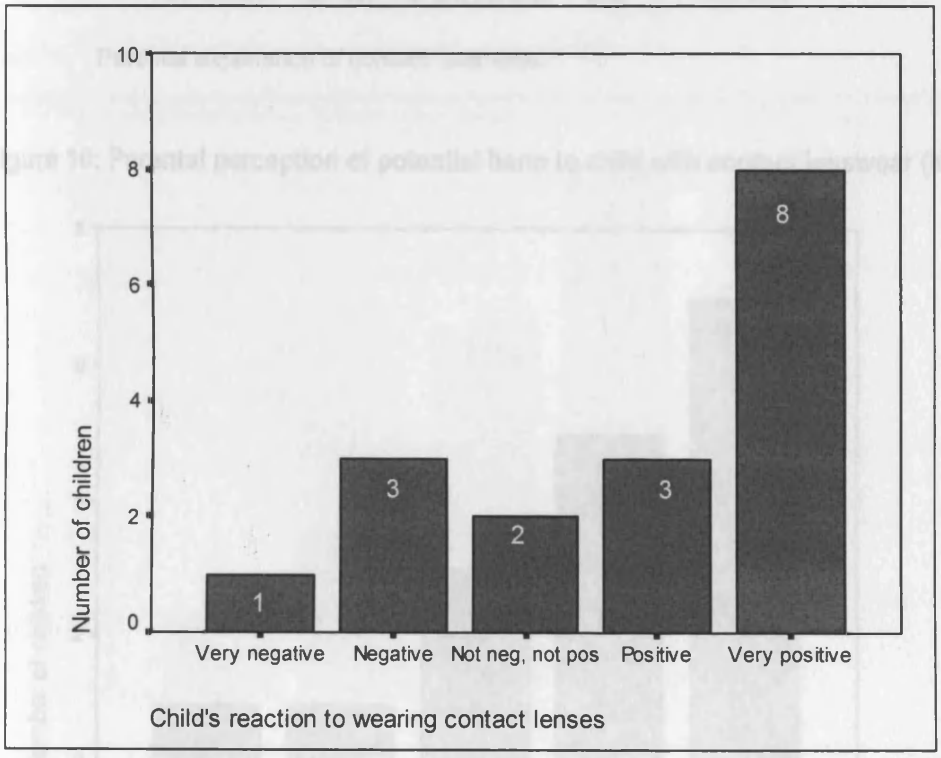


Figure 9: Parental experience of child's contact lens wear N=16 (6 not known)

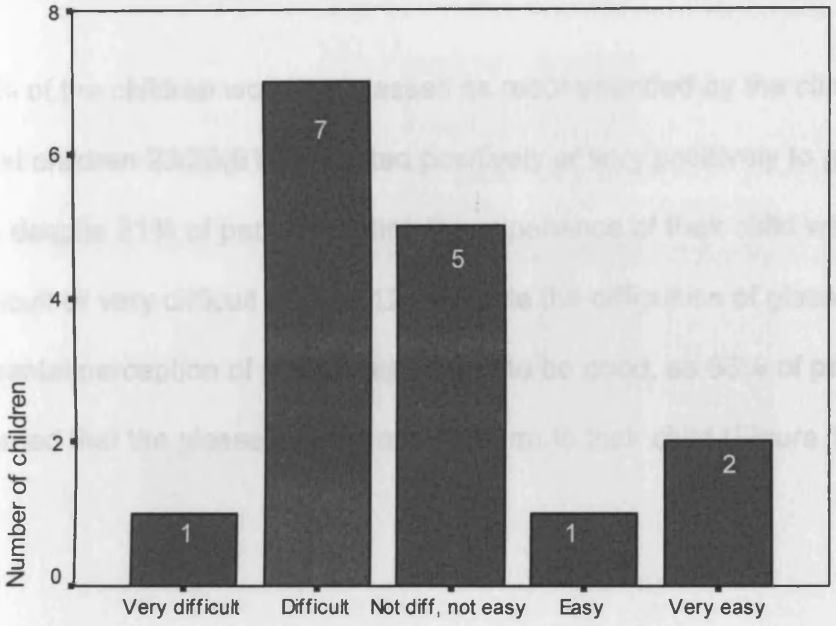
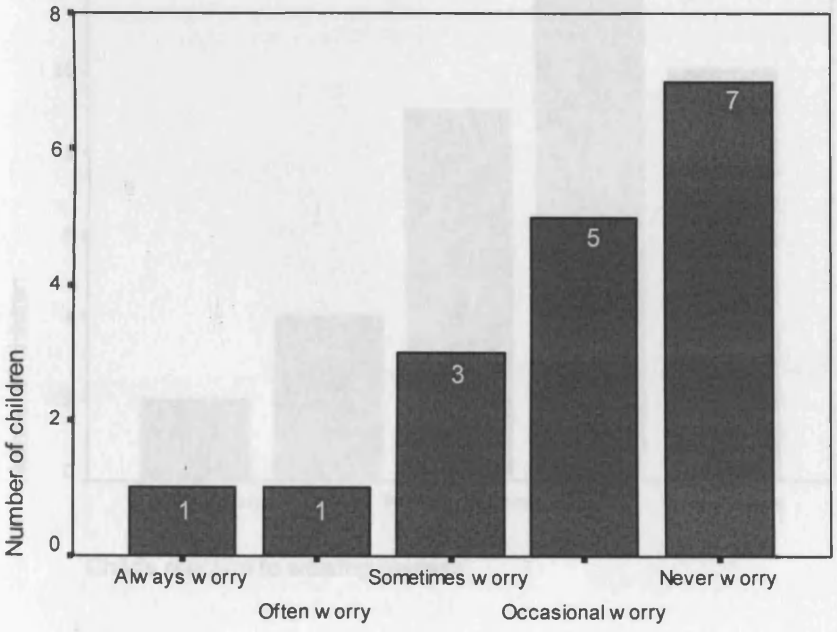


Figure 10: Parental perception of potential harm to child with contact lenswear (N=17)



5.2.2.3 Glasses

84% of the children wore the glasses as recommended by the clinicians.

Most children 23/38(61%) reacted positively or very positively to glasses (Figure 11) despite 21% of parents finding the experience of their child wearing glasses difficult or very difficult (Figure 12). Despite the difficulties of glasses wear, parental perception of glasses appeared to be good, as 68% of parents never worried that the glasses were causing harm to their child (Figure 13)

Figure 11: Child's reaction to wearing glasses (N=38)

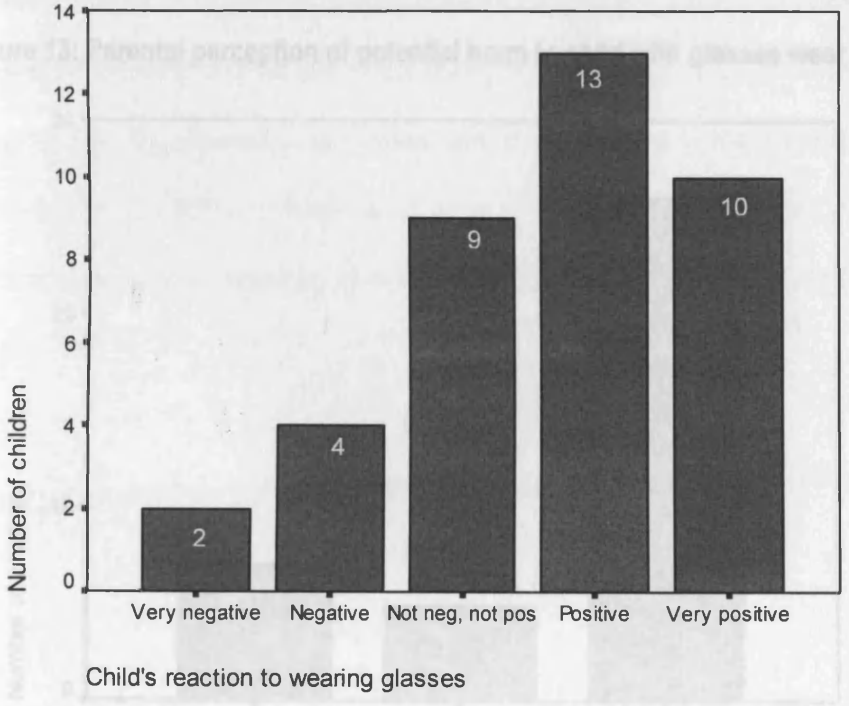
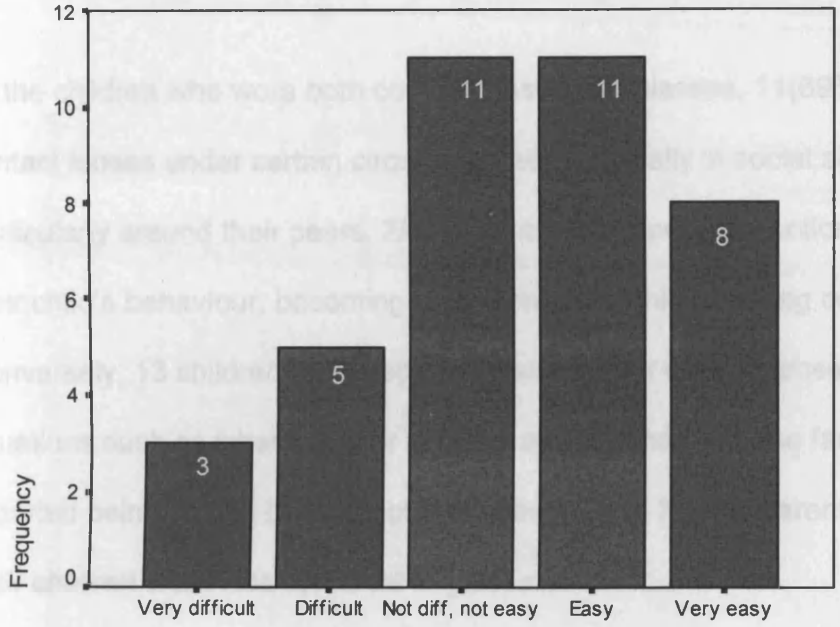
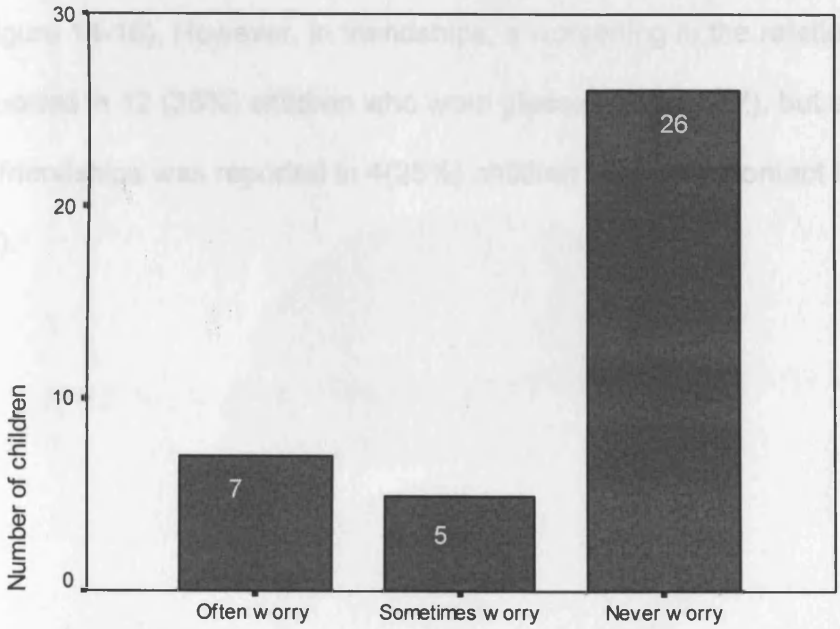


Figure 12: Parental experience of child's glasses wear (N=38)



Parental experience of contact lens wear

Figure 13: Parental perception of potential harm to child with glasses wear (N=38)



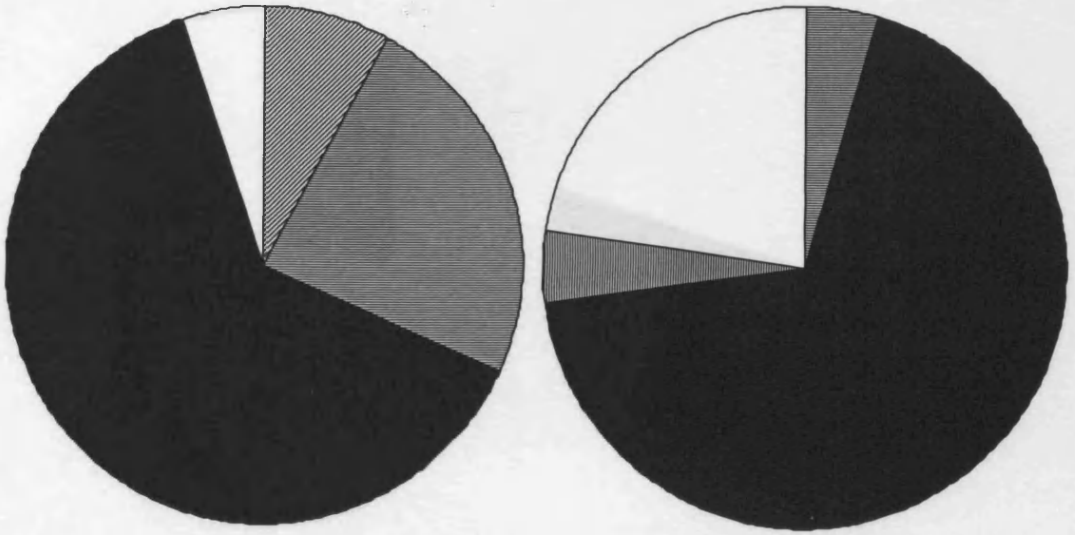
Parental perception of potential harm to child with glasses wear

5.2.2.4 Effect of glasses and contact lenses on social interactions and relationships

Of the children who wore both contact lenses and glasses, 11(69%) preferred contact lenses under certain circumstances, especially in social situations particularly around their peers. 2/41 parents also reported a noticeable change in their child's behaviour, becoming more confident whilst wearing contact lenses. Conversely, 13 children preferred their glasses over contact lenses in other situations such as when tired, or relaxing at weekends with the family. 4 children reported being bullied because of their glasses and 2 other parents had noticed their children were less confident in glasses.

In the majority of children, use of contact lenses or glasses did not affect the relationships of the child and their parents, siblings and other family members (Figure 14-16). However, in friendships, a worsening in the relationship was reported in 12 (33%) children who wore glasses (Figure 17), but an improvement in friendships was reported in 4(25%) children who wore contact lenses (Figure 17).

Figure 14: Effect of use of contact lenses or glasses on child/parent relationship



Glasses

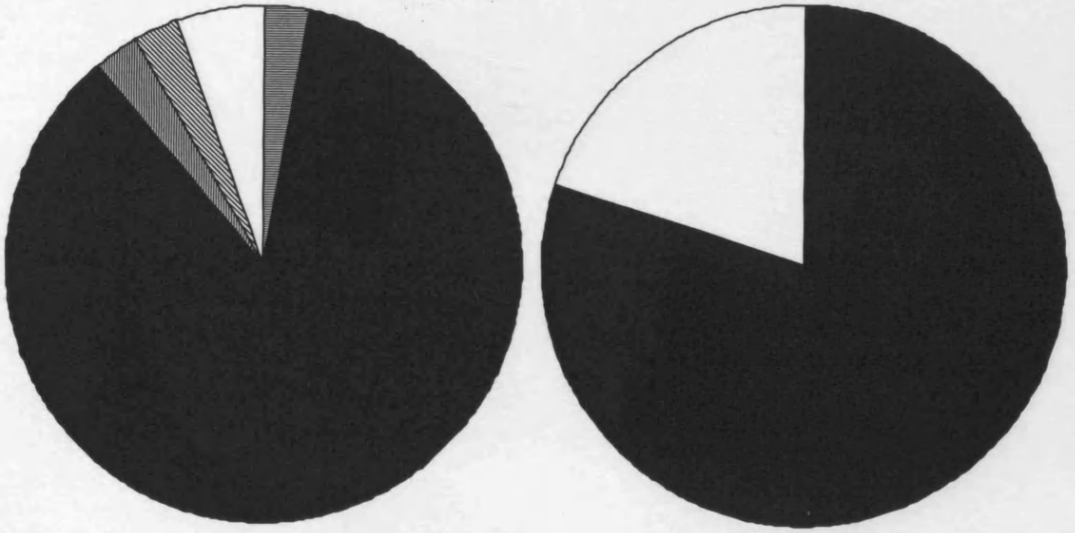
Contact lenses

	Glasses (N=38)	CL (N=20)
A lot worse	N=1	N=0
A little worse	N=0	N=1
No change	N=35	N=15
Improved a little	N=0	N=1
Improved a lot	N=1	N=0
Not known	N=1	N=4

20 children did not wear contact lenses, 1 child wore glasses, but was too young to discern any change in relationship with parent

3 children did not wear glasses

Figure 15: Effect of use of contact lenses or glasses on child/sibling relationship



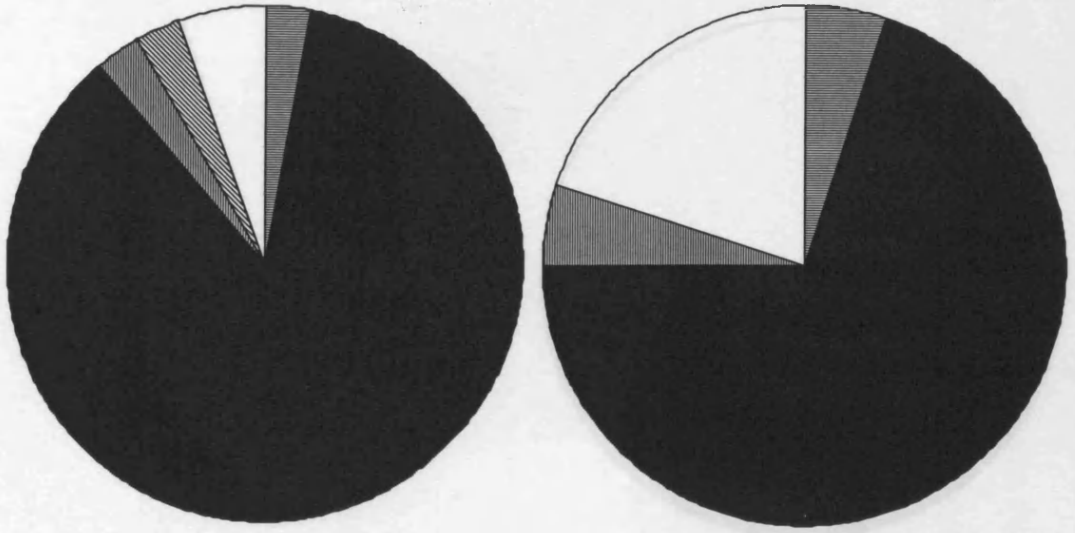
Glasses Contact lenses

	Glasses (N=36)	CL (N=20)
A lot worse	N=0	N=0
A little worse	N=1	N=0
No change	N=31	N=16
Improved a little	N=1	N=0
Improved a lot	N=1	N=0
Not known	N=2	N=4

20 children did not wear contact lenses, 1 child wore contact lenses but had no interactions with siblings

3 children did not wear glasses, 2 child wore glasses but had no interactions with siblings

Figure 16: Effect of use of contact lenses or glasses on child/other family member relationship



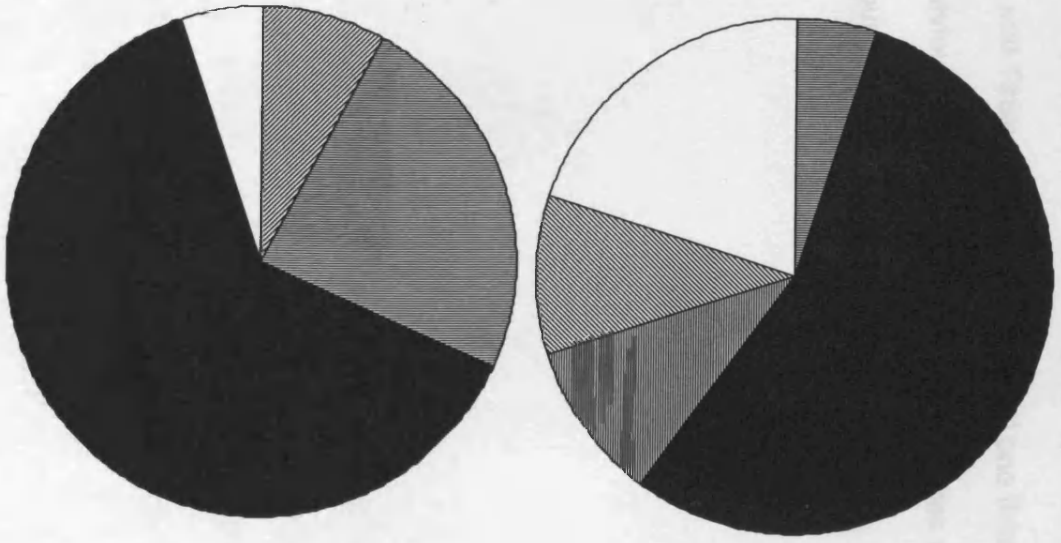
Glasses Contact lenses

	Glasses (N=35)	CL (N=20)
A lot worse	N=0	N=0
A little worse	N=0	N=1
No change	N=32	N=14
Improved a little	N=0	N=1
Improved a lot	N=0	N=0
Not known	N=3	N=4

20 children did not wear contact lenses, 1 child wore contact lenses but had no interactions with other family members

3 children did not wear glasses, 3 child wore glasses but had no interactions with other family members

Figure 17: Effect of use of contact lenses or glasses on child/friend relationship



Glasses

Contact lenses

	Glasses (N=38)	CL (N=20)
A lot worse	N=3	N=0
A little worse	N=9	N=1
No change	N=24	N=11
Improved a little	N=0	N=2
Improved a lot	N=0	N=2
Not known	N=2	N=4

20 children did not wear contact lenses, 1 child wore contact lenses but had no interactions with friends

3 children did not wear glasses

5.2.2.5 Occlusion

Of the 28/41 children who underwent or are undergoing occlusion, 19(67%) were girls. Median age at time of questioning was 6.99 years(6.25-12.19 years). Median age at commencement of occlusion (as recalled by parents) was 1 year(3 months to 6 years).

Just over a half of the parents (55.6%) managed to patch according to clinician's recommendations and Table 17 shows that almost one third (32%) of children changed their behaviour whilst being patched. All changes were a worsening or distressed behaviour.

Table 17: Change in child's behaviour attributed by parents to occlusion (N=28)

	Frequency	Percent
No change in behaviour	19	67.9
Readily upset	1	3.6
Less confident and more emotional	1	3.6
Miserable, bad tempered, withdrawn	1	3.6
Hides away when wearing patch	1	3.6
Occasional frustrated outburst	1	3.6
Became distressed and angry	1	3.6
Refused to wear patches	1	3.6
Hated occlusion	1	3.6
Not known	1	3.6

Most children (73%) reacted negatively or very negatively to occlusion (Figure 18) and likewise, 81% of parents found the experience of occlusion difficult or very difficult (Figure 19). Despite the difficulties of occlusion, parental perception of appeared to be good, as 63% of parents never worried that the patches were causing harm to their child (Figure 20).

Figure 18: Child's reaction to wearing patches (N=26)

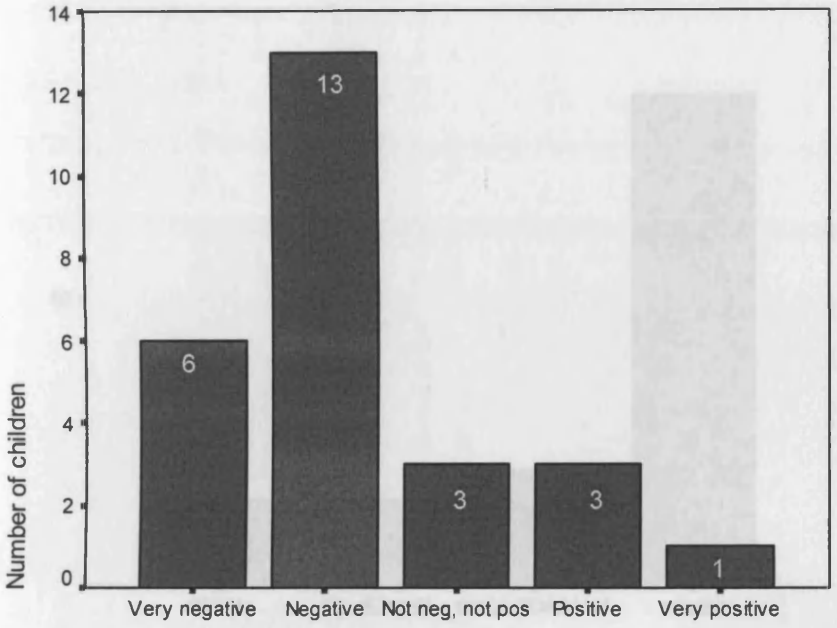


Figure 19: Parental experience of occlusion (N=26)

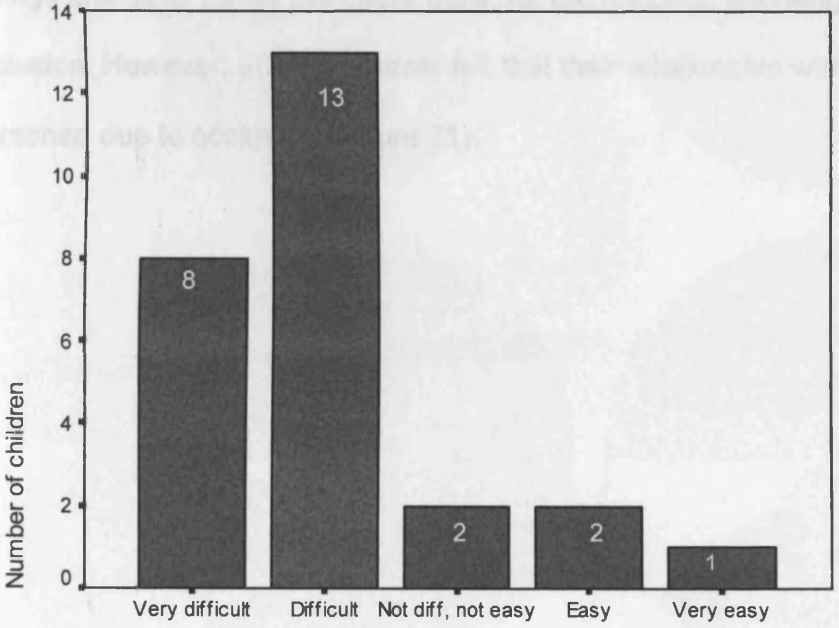
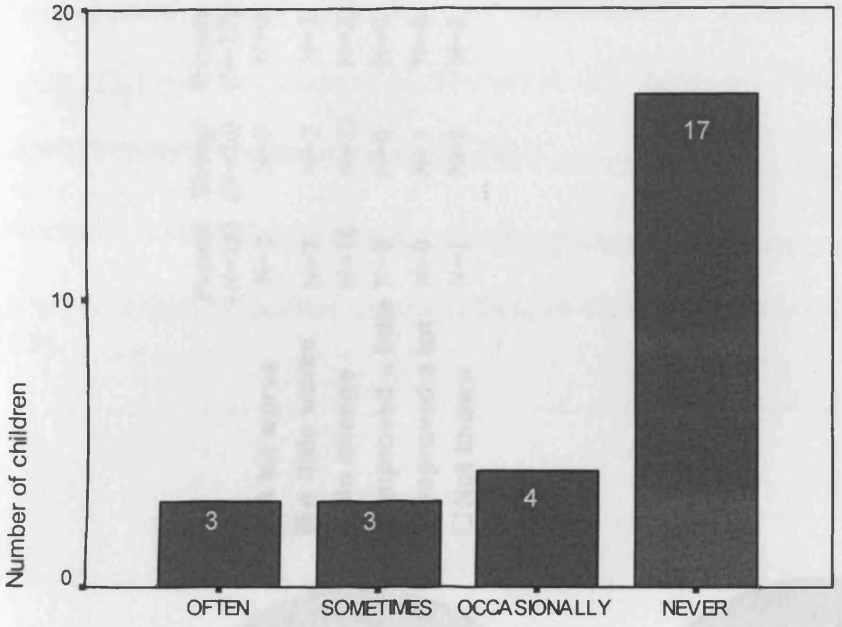
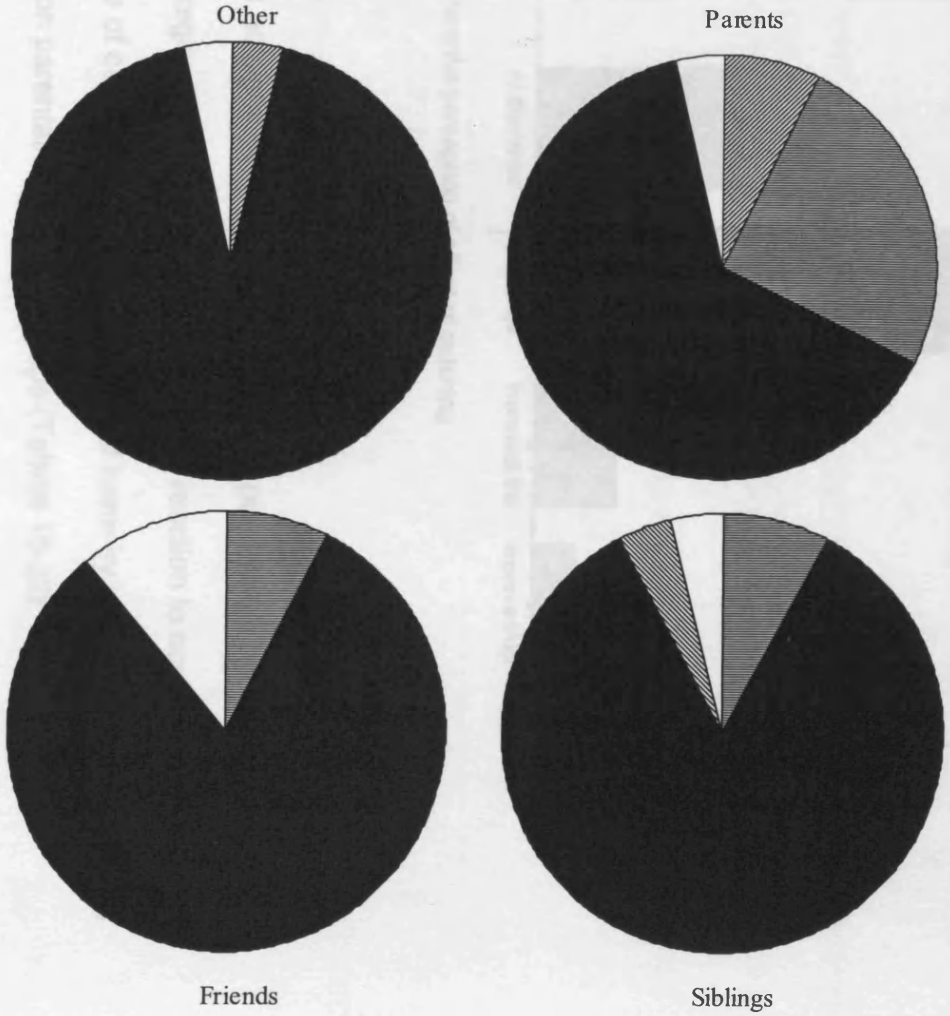


Figure 20: Parental perception of potential harm to child by occlusion(N=27)



Overall, the effect of occlusion on the child's relationship with their friends, siblings and other family members were, for the majority, unchanged by occlusion. However, 9(33%) parents felt that their relationship with their child had worsened due to occlusion (Figure 21).

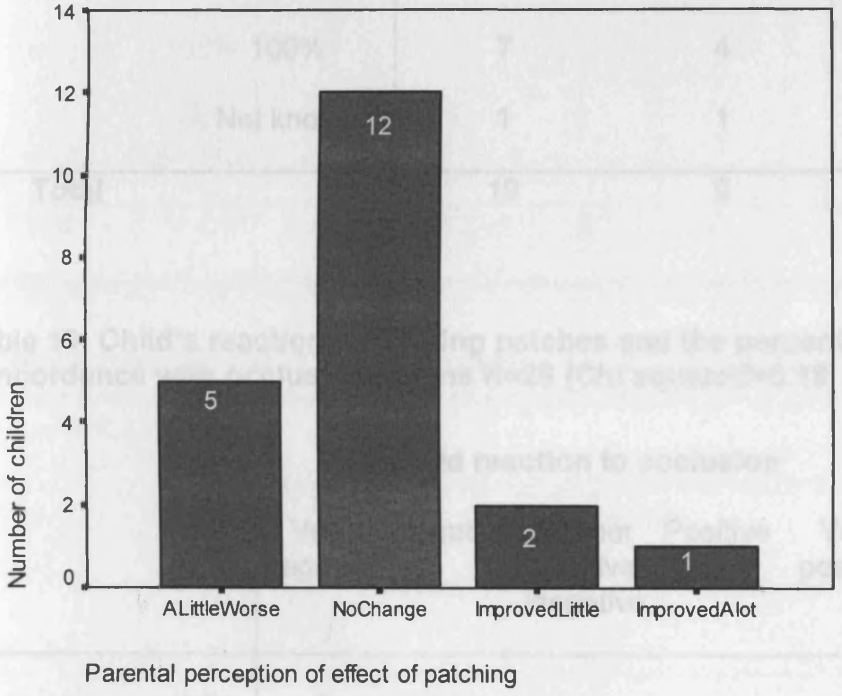
Figure 21: Effect of occlusion on the child's relationship with parents, siblings, friends and other family members



	Parents (N=28)	Siblings (N=26)	Friends (N=27)	Other (N=27)
A lot worse	N=2	N=0	N=0	N=1
A little worse	N=7	N=2	N=2	N=0
No change	N=18	N=22	N=22	N=25
Improved a little	N=0	N=0	N=0	N=0
Improved a lot	N=0	N=1	N=0	N=0
Not known	N=1	N=1	N=3	N=1

The parental expectations of the child's performance at school appeared in the majority of children to be unaffected by occlusion (60%), although 5 thought occlusion had worsened school performance, and 3 thought that occlusion had improved school performance (Figure 22)

Figure 22: Effect of occlusion on expected child performance at school (N=20)



There was no significant association between percentage concordance with the occlusion regime and the child's behaviour or reaction to occlusion, parental experience of occlusion, parental concern of harm by occlusion or the effect of occlusion on parental/child relationships (Tables 18-22)

Table 18: Change in child's behaviour attributed by parents to occlusion and the percentage concordance with occlusion regime N=28 (Chi-squared=1.39, p=0.71)

		Child behavioural change		Total
		No	Yes	
Percentage concordance with occlusion regime	<50%	5	2	7
	50%	2	0	2
	75%	4	2	6
	100%	7	4	11
	Not known	1	1	2
Total		19	9	28

Table 19: Child's reaction to wearing patches and the percentage concordance with occlusion regime N=26 (Chi squared=5.18, p=0.52)

		Child reaction to occlusion					Total
		Very negative	Negative	Neither positive /negative	Positive	Very positive	
Percentage concordance with occlusion regime	<50%	2	4			1	7
	50%		1		1		2
	75%	2	3	1			6
	100%	2	3	2	2		9
	Not known		2				2
Total		6	13	3	3	1	26

Table 20: Parental experience of occlusion and the percentage concordance with occlusion regime N=26 (Chi-squared=5.18, p=0.52)

		Parental experience of occlusion					Total
		Very difficult	Difficult	Neither difficult nor easy	Easy	Very easy	
Percentage concordance with occlusion regime	<50%	2	4			1	7
	50%	1			1		2
	75%	3	3				6
	100%	2	5	1	1		9
	Not known		1	1			2
Total		8	13	2	2	1	26

Table 21: Parental perception of potential harm to child by occlusion and the percentage concordance with occlusion regime N=27 (Chi-squared=5.13, p=0.53)

		Parental perception of potential harm to child by occlusion					Total
		Very often	Often	Sometimes	Occasionally	Never	
Percentage concordance with occlusion regime	<50%		1	1	1	4	7
	50%		1		1		2
	75%			1		5	6
	100%		1	1	2	6	10
	Not known					2	2
Total			3	3	4	17	27

Table 22: Change in the relationship between child and their parents due to occlusion and the percentage concordance with occlusion regime N=27 (Chi-squared=1.57, p=0.67)

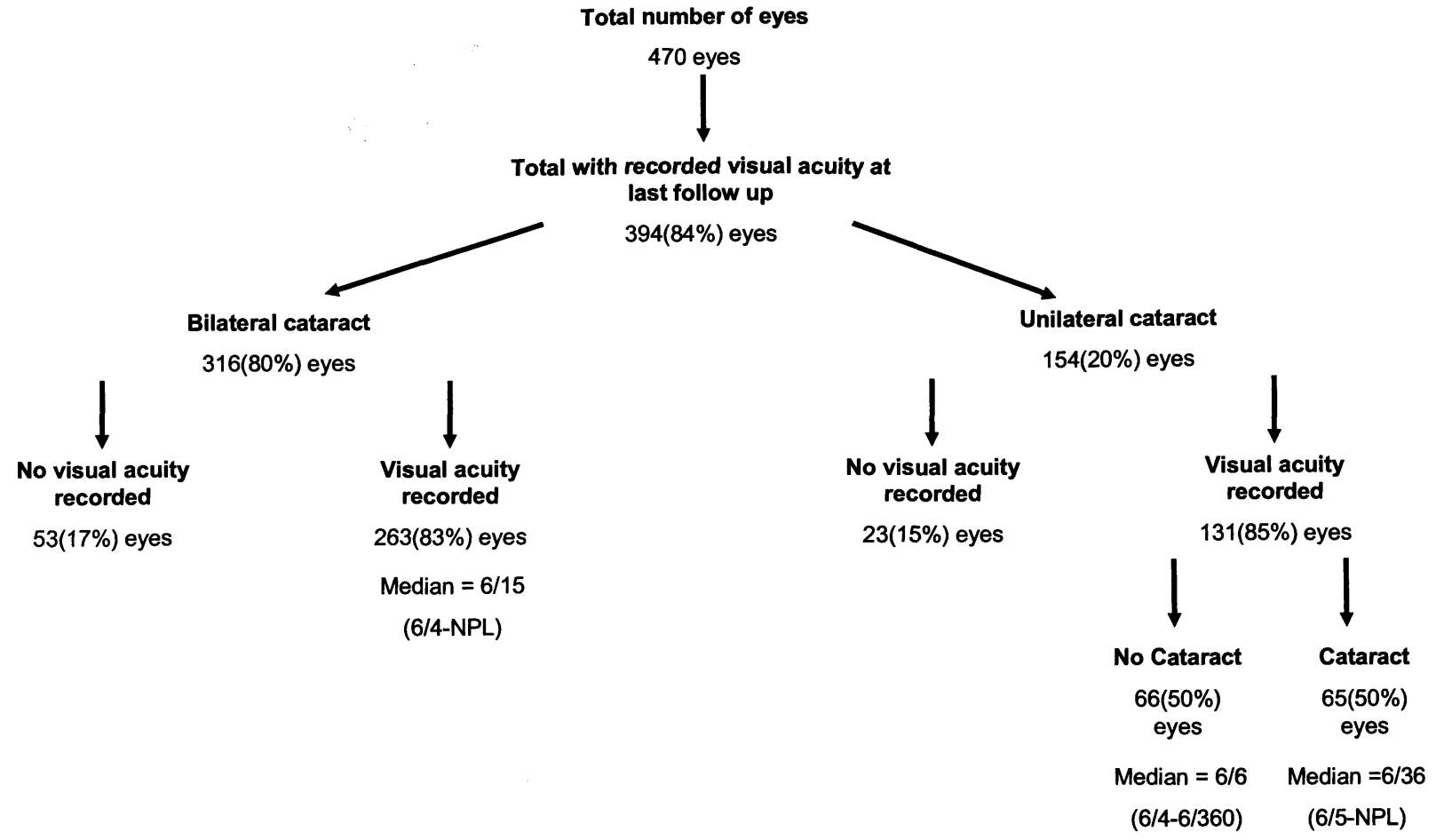
		Change in the relationship between child and their parents due to occlusion					Total
		A lot worse	A little worse	No change	A little better	A lot better	
Percentage concordance with occlusion regime	<50%	1	1	5			7
	50%			2			2
	75%	1	1	4			6
	100%		4	6			10
	Not known		1	1			2
Total		2	7	18			27

5.3 Outcomes of children with congenital cataract

5.3.1 Visual acuity

At the point of follow up for this study, visual acuity measurements were available for 394 (84%) of all eyes as shown in Figure 23. These were Snellen measures in 323(82%) eyes. Data for 71(18%) eyes which were recorded as logMAR visual acuity were transformed to Snellen notation for analysis, as discussed earlier (*refer to section 4.3.2.2 Visual acuity (VA), page 81*). No visual acuity was recorded in 78 eyes either due to difficulties with examination of children with learning difficulties or failure to record the visual acuity in the questionnaire.

Figure 23: Visual acuity recordings at final follow up



For the purposes of analysis of visual acuity, 12 children aged <1825 days (5 years) old at follow up examination were excluded. This was to ensure that only children with reliable and stable acuity measures were included and all children were at an age at which most postoperative complications would have manifest. Notably, the median and range of visual acuities of the children in the excluded group were similar to those aged >5 years old (as shown in Figures 24-26, Appendix 4).

Figures 27-29 show the distribution of the visual acuities. Of the eyes that underwent surgery, the median visual acuity of children with bilateral cataract, by eye was 6/19 (range=6/5 to PL) and in unilateral cataracts the median was 6/48 (range=6/5 to NPL). Of the eyes of children with bilateral cataract that did not undergo surgery, the median visual acuity was 6/12 (range=6/4 to NPL) and in unilateral cataracts the median was 6/24 (range=6/5 to PL). Of the non-cataractous eyes of children with unilateral cataract, the median visual acuity was 6/6 (range=6/4 to 6/360).

Figure 27: Visual acuity of children with bilateral cataract aged >5 years at follow up (N=246 eyes)

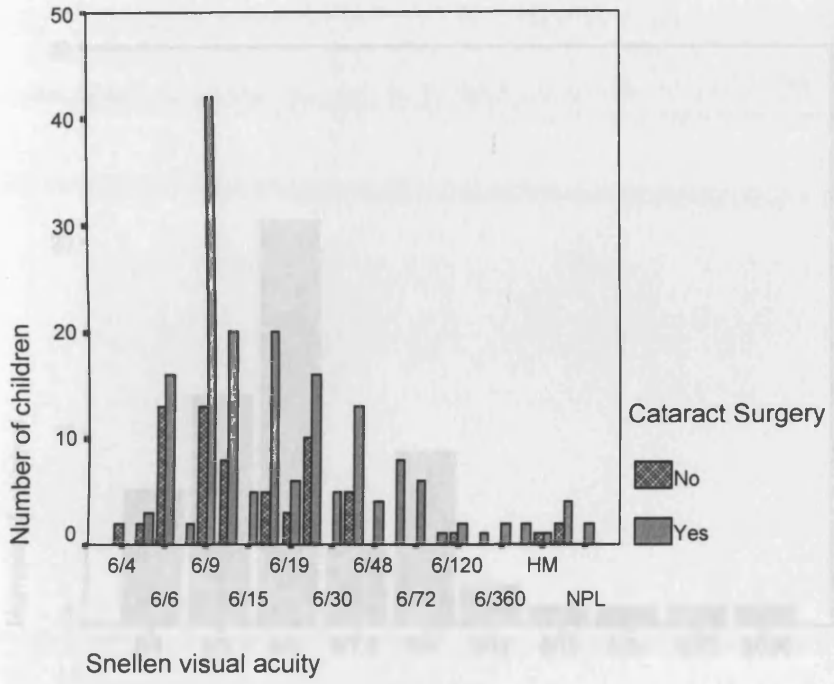


Figure 28: Visual acuity of cataractous eyes of children with unilateral cataract aged>5 years at follow up (N=58 eyes)

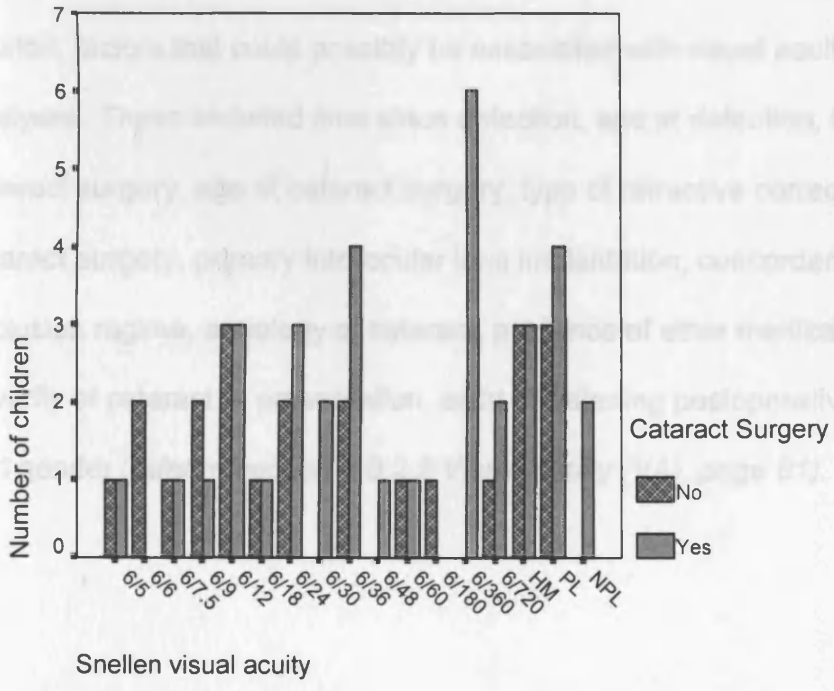
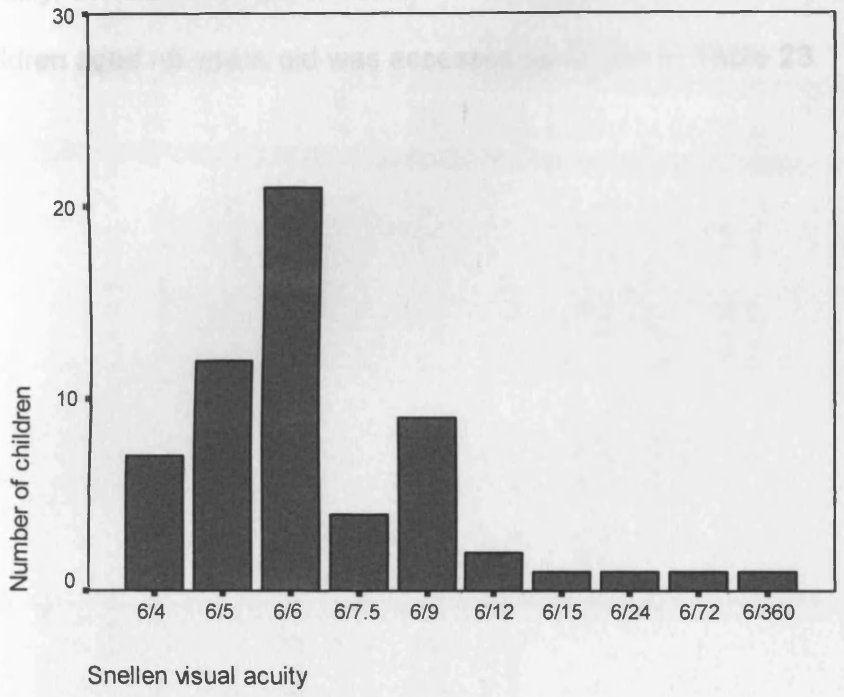


Figure 29: Visual acuity of non-cataractous eyes of children with unilateral cataract at follow up (N=59 eyes)



A priori, factors that could possibly be associated with visual acuity were analysed. These included time since detection, age at detection, time since cataract surgery, age at cataract surgery, type of refractive correction, type of cataract surgery, primary intraocular lens implantation, concordance with occlusion regime, aetiology of cataract, presence of other medical conditions, severity of cataract at presentation, sight threatening postoperative complications and gender (*refer to section 4.3.2.2 Visual acuity (VA), page 81).*

5.3.1.1 Factors affecting the visual acuity of children with bilateral cataracts

Initially, correlation between factors of interest in relation to visual acuity of children aged >5 years old was assessed as shown in Table 23.

Table 23: Association between the factors of interest in relation to visual acuity of children aged >5 years old with bilateral cataracts

α=Spearman's correlation coefficient, β=Kruskal-Wallis score, γ=Mann-Whitney U Score and δ=Chi squared

	Time detect	Age detect	Time surg	Surg age	Correct	Surg type	Patching	Aetiol	Other meds	Gender	Severe Cat	IOL	Complication
Time since detection	-0.18 α p=0.00	0.65 α p=0.00	-0.10α p=0.16	0.15β p=0.93	6.81β p=0.03	7.56β p=0.06	19.70β p=0.00	4.05γ p=0.00	-0.29γ p=0.77	-1.44γ p=0.15	-0.84γ p=0.40	-1.71γ p=0.09
Age at detection		-0.28 α p=0.00	0.77α p=0.00	11.31β p=0.00	28.80β p=0.00	5.25β p=0.16	11.64β p=0.01	-0.93γ p=0.35	-1.70γ p=0.09	-3.67 γ p=0.00	-7.54γ p=0.00	-2.82γ p=0.01
Time since cataract surgery			-0.56α p=0.00	3.16β p=0.21	16.73β p=0.00	7.22β p=0.07	5.98β p=0.11	0.10γ p=0.92	-1.80 γ p=0.07	-5.30γ p=0.00	-5.65γ p=0.00	-1.96γ p=0.05
Age at cataract surgery				9.01β p=0.01	58.42β p=0.00	8.39β p=0.04	6.73β p=0.08	-2.88γ p=0.00	-0.82γ p=0.41	-5.28γ p=0.00	-10.24γ p=0.00	-3.04γ p=0.00
Type of refractive correction					16.50δ p=0.04	3.94δ p=0.69	4.87δ p=0.56	3.45δ p=0.18	6.48 δ p=0.04	12.94δ p=0.00	22.24δ p=0.00	1.09δ p=0.58
Type of cataract surgery						7.18δ p=0.10	18.68δ p=0.00	42.26δ p=0.05	9.36δ p=0.20	5.92δ p=0.00	169.91δ p=0.00	65.38δ p=0.07
Patching compliance							28.62δ p=0.00	2.34δ p=0.50	8.29δ p=0.04	1.46δ p=0.69	16.74 δ p=0.00	2.13δ p=0.55
Aetiology of cataract								163.70δ p=0.00	2.92δ p=0.40	22.70δ p=0.00	14.22 δ p=0.00	12.48δ p=0.01
Presence of other medical disorders									1.80δ p=0.77	0.90δ p=0.14	2.16δ p=0.97	0.00δ p=0.1
Gender										3.08δ p=0.08	3..53δ p=0.06	1.03δ p=0.31
Severe cataract at detection											36..35δ p=0.00	6.30δ p=0.01
Postoperative complication												1.80δ p=1.08

There was a significant positive correlation between age at detection and age at surgery (0.77, p=0.00), as earlier detection would allow earlier surgery. Age of detection and age of surgery were also highly associated with surgical techniques (type of surgery (28.80 p=0.00; 58.42, p=0.00), and IOL implantation (-7.54, p=0.00;U=-10.24, p=0.00)). This may be because the age of an infant may determine the type of surgery, e.g. a very young infant probably would not have an IOL implantation, and so lensectomy/vitrectomy would be appropriate. Age at surgery was also associated with postoperative sight threatening complications (-3.04, p=0.00).

Age at detection was associated with a severe cataract at detection (-5.28, 0.00) and aetiology of the cataract (11.64, p=0.01) as more severe cataract may be more easily noticed and certain aetiology, for example in association with other disorders may be brought to the attention of clinicians earlier.

Occlusion concordance was associated with the age at surgery (8.39, p=0.04), as children at different ages may differ in their levels of cooperation with their parents.

Factors of interest were assessed univariately in relation to visual acuity of children aged >5 years old and the distribution of these are shown in Figures 30 to 42, Appendix 5.

Using ordinal logistic regression, the relationships between visual acuity and the variables of interest were analysed univariately whose results are shown inTable 24 for children with bilateral cataracts. Factors which were statistically significant at the 0.05 level are indicated in bold.

Table 24: Univariate ordinal logistic regression analysis of visual acuity of 182 eyes of 98 children with bilateral cataract, taking into account clustering within child

Variable	Relative odds of having worse acuity (CI)	P value	Clustering within child 95% CI (p value)
Time since detection (days)	1.00(0.99 to 1.00)	0.67	0.99 to 1.00 (0.74)
Age at detection(days)	**0.999(0.99 to 0.99)**	**0.00**	**0.99 to 0.99 (0.00)**
Time since surgery (days)	1.00(0.99 to 1.00)	0.14	0.99 to 1.00 (0.18)
Age at cataract surgery (days)	**0.99(0.99 to 0.99)**	**0.00**	**0.99 to 0.99 (0.00)**
Correction (Glasses) *(baseline)			
Correction (CL)*	1.36(0.66 to 2.84)	0.47	0.57 to 3.41 (0.49)
Correction (CL and Glasses)*	**8.53(1.46 to 49.80)**	**0.02**	**1.37 to 51.71 (0.05)**
Type of cataract surgery (Aspiration alone)#(baseline)			
Type of cataract surgery (Lens aspiration and vitrectomy)#	**0.34(0.19 to 0.83)**	**0.01**	**0.10 to 0.92 (0.02)**
Type of cataract surgery (Lensectomy-vitrectomy)#	**3.55(1.90 to 6.62)**	**0.00**	**1.67 to 7.12 (0.00)**
Primary IOL implantation	**0.45 (0.26 to 0.76)**	**0.00**	**0.22 to 0.91 (0.03)**
100% concordance with occlusion+(baseline)			
50% concordance with occlusion+	0.34(0.10 to 1.19)	0.09	0.08 to1.51 (0.156)
<50% concordance with occlusion regime+	2.18(0.93 to 6.67)	0.07	0.58 to 6.87 (0.07)
Aetiology (Systemic)∞ (baseline)			
Aetiology (Isolated)∞	**0.15(0.07 to 0.31)**	**0.00**	**0.05 to 0.47 (0.00)**
Aetiology (Ocular) ∞	0.53(0.23 to 1.21)	0.134	0.16 to 1.78 (0.30)
Aetiology(Ocularsystemic)∞	0.30(0.02 to 3.70)	0.350	0.02 to 3.92 (0.57)
Presence of other medical conditions	**2.28(1.30 to 4.02)**	**0.00**	**1.11 to 4.81 (0.03)**
Severe cataract at presentation	1.56(0.97 to 2.49)	0.065	0.92 to 2.85 (0.09)
Presence of sight threatening postoperative complication	**2.09(1.08 to 4.03)**	**0.03**	**0.91 to 4.79 (0.08)**
Gender	1.49(0.96 to 2.30)	0.08	0.86 to 2.56 (0.16)

Correction – (CL and Glasses vs CL) (Relative odds=0.16 CI=0.02 to 1.03, p=0.054) clustering within child: CI=0.04 to 7.09 p=0.34

Type of cataract surgery – (Lens aspiration and vitrectomy vs Lensectomy-vitrectomy) (Relative odds=1.40 CI=0.71 to 1.74, p=0.46) clustering within child:0.57 to 3.42 p=0.46

+100 vs 75% concordance with occlusion regime was insignificant (Relative odds =1.62 CI=0.65 to3.44, p=0.34) clustering within child:0.71 to 3.53 (0.27)

∞Aetiology- (Ocular vs Isolated) was significant (Relative odds =0.29 CI=0.15 to 0.52, p=0.00) clustering within child:0.15 to 0.55, p=0.00

Aetiology- (Ocular vs ocularsystemic) was significant (Relative odds =0.55 CI=0.05 to 6.81, p=0.66) clustering within child: 0.33 to 1.11, p=0.05

Aetiology- (Ocularsystemic vs isolated) was insignificant (Relative odds =0.50 CI=0.04 to 5.69,p=0.576) clustering within child:0.35 to0.70, p=0.00

In the univariate analysis, the associations between visual acuity and age at detection of the cataract, age of cataract surgery, type of refractive correction, type of cataract surgery, primary intraocular lens implantation, aetiology of the cataract, presence of other medical conditions, and sight threatening complications were statistically significant at the 0.05 level. As expected, clustering within child produced the same relative odds, but with a widening of the confidence interval (and a possible decrease in significance).

As discussed earlier in the methods *(refer to section 4.3.2.2 Visual acuity (VA), page 81)* and shown in Table 25, the multivariate regression model was constructed with variables significant at the 0.05 level using ordinal regression.

Table 25: Multivariate ordinal regression model of factors associated with visual acuity at follow up examination of 182 eyes of 98 children with bilateral cataracts

Variable	Relative odds of having worse acuity(CI)	P value	Clustering within child 95% CI (p value)
Age at cataract surgery (days)	1.00(1.00 to 1.00)	0.01	0.99 to 1.00 (0.018)
100% concordance with occlusion regime+ (baseline)			
50% concordance with occlusion regime+	5.64(1.51 to 21.05)	0.01	0.05 to 0.81 (0.02)
<50% concordance with occlusion regime+	1.17(0.27 to 5.03)	0.83	0.26 to 5.18 (0.84)
Presence of other medical conditions	3.53(1.20 to 10.41)	0.02	1.08 to11.44 (0.04)
Presence of sight threatening postoperative complications	2.94(1.09 to 7.88)	0.03	1.38 t 6.51 (0.04)
Severe cataract at presentation	0.06(0.28 to 0.01)	0.00	0.01 to 0.26 (0.01)

+100vs75%: Relative odds=0.56, CI=0.24 to 1.32 p=0.18 clustering within child=0.22 to 1.42, p=0.22

In the multivariate model, the factors from the univariate analysis that remained significant at the 0.05 level were age at cataract surgery, presence of other medical conditions and presence of sight threatening complications. The level of concordance with occlusion regime, and severity of cataract at presentation became significant in the multivariate model, having not been significant in the univariate model.

The analysis suggested that the odds of being in a worse visual acuity category increased with increasing age at surgery, being 1.09 at 3 months compared to 1.43 at 1 year (ie children operated on earlier have a better visual acuity). The odds of being in a worse visual acuity category were 3.53 times greater for those with additional medical condition(s) compared to those without. The odds of being in a worse visual acuity category were 2.94 times greater for those with a sight threatening postoperative complication than those without. In addition, the odds of being in the next worse visual acuity category were 5.64 for those achieving <50% concordance with their occlusion occlusion regime compared to those achieving 100% concordance. Finally, the odds of being in a worse visual acuity category were 0.06 for those with a severe cataract at presentation than those without, which contradicts the findings of the univariate analysis in which those with a severe cataract at presentation had a worse visual acuity (odds 1.56).

5.3.1.2 Factors affecting the visual acuity of children with unilateral cataracts

Outcomes of eyes with and without cataract in children with unilateral cataracts would be expected to be very different (*refer to section 2.2.2 Management of congenital cataract by the laterality of the cataract, page 34)* and therefore only cataractous eyes were analysed.

Initially, correlation between factors of interest in relation to visual acuity of children aged >5 years old was assessed as shown in Table 26.

Table 26: Association between the factors of interest in relation to visual acuity of children aged >5 years old with unilateral cataracts (eyes with cataract only)

(α=Spearman's correlation coefficient, β=Kruskal-Wallis score, γ=Mann-Whitney U Score and δ=Chi squared)

	Time Detect	Age Detect	Time Surg	Surg Age	Correct	Surg Type	Patching	Aetiol	Other Meds	Gender	Severe Cat	IOL	Complic
Time Since Detection		-2.07 α p=0.024	0.98 α p=0.00	-0.15α p=0.39	0.82β p=0.37	1.60β p=0.45	0.75β p=0.86	0.46β p=0.93	-0.17γ p=0.86	-2.00 γ p=0.04	-0.04γ p=0.97	-0.37γ p=0.71	-0.56γ p=0.57
Age at Detection			-0.19 α p=0.27	0.89α p=0.00	11.43β p=0.00	1.01β p=0.01	10.04β p=0.02	1.72β p=0.63	0.65γ p=0.52	-0.50γ p=0.62	-0.95 γ p=0.34	-4.33γ p=0.00	-0.73γ p=0.47
Time Since Cataract Surgery				-0.28α p=0.08	0.05β p=0.83	0.21β p=0.90	0.47β p=0.93	4.93β p=0.18	-0.87γ p=0.38	-0.79 γ p=0.43	-1.15γ p=0.25	-0.49γ p=0.63	-0.31γ p=0.76
Age at Cataract Surgery					10.62β p=0.00	9.88β p=0.01	5.48β p=0.14	2.06β p=0.56	-0.27γ p=0.79	-0.64γ p=0.52	-1.97γ p=0.05	-3.90γ p=0.00	-1.67γ p=0.10
Type of Refractive Correction						13.70δ p=0.00	7.15δ p=0.07	3.84δ p=0.15	0.70δ p=0.40	0.33 δ p=0.56	2.90δ p=0.09	12.98δ p=0.00	0.01δ p=0.92
Type of Cataract Surgery							16.13δ p=0.06	14.35δ p=0.11	5.25δ p=0.16	4.67δ p=0.20	9.53δ p=0.02	9.97δ p=0.01	4.42δ p=0.11
Patching Compliance								11.11δ p=0.26	1.76δ p=0.62	1.23δ p=0.75	3.13δ p=0.37	6.35δ p=0.10	5.43δ p=0.14
Aetiology of cataract									28.97δ p=0.00	2.14δ p=0.49	0.49δ p=0.92	4.54 δ p=0.21	4.02δ p=0.26
Presence of other medical disorders										p=0.08	3.03δ p=0.62	0.25δ p=0.88	0.02δ p=0.37
Gender											0.96δ p=0.33	6.64δ p=0.01	4.09δ p=0.30
Severe cataract at presentation												0.25δ p=0.62	0.01δ p=0.91
Presence of postoperative complication													p=0.68

There was a significant positive correlation between age at detection and age at surgery (0.89, p=0.00), as earlier detection would allow earlier surgery. Age of detection and age of surgery were also highly associated with surgical techniques (type of surgery (1.01 p=0.01; 9.88, p=0.01), and IOL implantation (-4.33, p=0.00;U=-3.90, p=0.00)). This may be because the age of an infant may determine the type of surgery, e.g. a very young infant probably would not have an IOL implantation, and so lensectomy/vitrectomy would be appropriate. Age at surgery was not associated with postoperative sight threatening complications (-1.67, p=0.10). As may be expected, occlusion concordance was associated with the age at detection (10.04, p=0.02), as children at different ages may differ in their levels of cooperation with their parents.

Factors of interest were assessed univariately in relation to visual acuity of children aged >5 years old and the distribution of these are shown in Figures 43 to 55, Appendix 6.

Using logistic ordinal regression, the relationships between visual acuity and variables of interest were analysed univariately for children with unilateral cataract whose results are shown in Table 27. Factors which were statistically significant at the 0.05 level are indicated in bold.

Table 27: Univariate ordinal regression analysis of 38 eyes (with cataract) of 38 children with unilateral cataract

Variable	Relative odds of having a worse acuity(CI)	P value
Time since detection (days)	1.00(0.999 to 1.00)	0.37
Age at detection(days)	**0.999(0.999 to 1.00)**	**0.05**
Time since surgery (days)	0.999(0.998 to 1.00)	0.41
Age at cataract surgery (days)	0.999(0.999 to 1.00)	0.32
Correction (Glasses) *(baseline)		
Correction (CL)*	2.22(0.53 to 9.21)	0.27
Type of cataract surgery (Lens aspiration alone)# (baseline)		
Type of cataract surgery (Lens aspiration and vitrectomy)#	0.99(0.21 to 4.73)	0.99
Type of cataract surgery (Lensectomy-vitrectomy)#	0.51(0.10 to 2.62)	0.42
Primary IOL implantation	**0.20(0.06 to 0.743)**	**0.02**
100% concordance with occlusion regime+(baseline)		
50% concordance with occlusion regime+	**10.18(1.60 to 64.72)**	**0.01**
<50% concordance with occlusion regime+	2.18(1.69 to 33.62)	0.08
Aetiology (OcularSystemic)∞ (baseline)		
Aetiology (Isolated)∞	0.21(0.02 to 2.69)	0.23
Aetiology (Ocular) ∞	0.33(0.03 to 4.06)	0.230
Presence of other medical conditions	1.00(0.29 to 3.47)	0.99
Severe cataract at presentation	**5.05(1.56 to 16.35)**	**0.01**
Presence of sight threatening postop complication	3.86(0.78 to 18.86)	0.10
Gender	0.64(0.26 to 1.59)	0.34

Type of cataract surgery - (Lens aspiration and vitrectomy vs Lensectomy-vitrectomy) (Relative odds=1.95 CI=2.62 to 7.28, p=0.32)

+100 vs 75% concordance with occlusion regime (Relative odds=0.89 CI=0.11 to 7.29, p=0.91)

∞Aetiology- (Ocular vs Isolated) (Relative odds=0.01CI=0.26 to 1.63, p=0.36)

In the univariate analysis, the associations between visual acuity and age of detection of the cataract, presence of a primary IOL implant, concordance with the occlusion regime and presence of a severe cataract at presentation were statistically significant at the 0.05 level.

Using ordinal logistic regression, a multivariate model was constructed whose output is shown in Table 28.

Table 28: Multivariate ordinal logistic regression model of factors associated with visual acuity at follow up examination of 38 cataractous eyes of 38 children with unilateral cataracts

Variable	Relative odds of having a given acuity or worse(Confidence interval)	P value
Age at detection (days)	0.999(0.99 to 1.00)	0.56
100% concordance with occlusion regime+		
50% concordance with occlusion regime+	**14.29(1.36 to 149.6)**	**0.03**
<50% concordance with occlusion regime+	**5.64(5.45 to 663.15)**	**0.00**
Severe cataract at presentation	0.34(8.15 x10^{-4} to14.53)	0.58
Primary IOL implantation	1.39(0.16 to 12.06)	0.77

+100 vs 75% concordance with occlusion regime (Relative odds=0.60 CI=0.06 to 5.58 p=0.652)

In the multivariate model, the only factor that remained significant at the 0.05 level was concordance with the occlusion regime. However, this is strongly associated with age at detection (Table 26). Age at detection determines timing of treatment, and as the mechanism of amblyopia is strongly dependent on timing of intervention (*refer to section 2.1.2 Visual Development, page 21*), this factor was kept in the multivariate model (Table 29).

Table 29: Multivariate ordinal regression model of factors associated with visual acuity at followup examination of 58 eyes (with cataract) of 58 children with unilateral cataracts

Variable	Relative odds of having a given acuity or worse(Confidence interval)	P value
Age at detection (days)	1.00(0.99 to 1.00)	0.59
100 vs <50% concordance with occlusion regime+	7.92(1.68 to 37.26)	0.01
100 vs 50% concordance with occlusion regime+	9.78(1.34 to 71.00)	0.024

+100 vs 75% concordance with occlusion regime (Relative odds =0.99 CI=0.11 to 7.81 p=0.95)

Thus the odds of being in the next worse visual acuity category were 7.92 for those achieving <50% concordance with their occlusion regime compared to those achieving 100% concordance. This increased to an odds of 9.78 for those achieving 50% concordance with their occlusion regime compared to those achieving 100% concordance (but was less significant). Having taken into account occlusion concordance, the odds of being in a worse visual acuity category if detected at 6 weeks was 1.010 compared to 1.096 detected at 1 year (ie children detected earlier have a better visual acuity).

5.3.2 Complications of cataract surgery

53 eyes had a sight threatening postoperative complication as shown in Table 30:

Table 30: Number of children with sight threatening complications

Complication	Laterality of cataract	
	Bilateral cataracts N=34 children, 44 eyes	**Unilateral N=9 children, 9 eyes**
Glaucoma (Postoperative open angle and closed angle)	18 children 27 eyes	6 children 6 eyes
Retinal detachment	2 children 3 eyes	1 child 1 eye
Endophthalmitis	1 child 1 eye	
Wound leak	1 child 1 eye	1 child 1 eye
Vitreous haemorrhage requiring vitrectomy	1 child 1 eye	
Vitreous prolapse (to AC/wound/leak) requiring vitrectomy	10 children 10 eyes	1 child 1 eye
Collapsed anterior chamber requiring revision	1 child 1 eye	

5.3.2.1 Glaucoma

Glaucoma occurred in 30(13%) of children with congenital cataract (Table 31). Glaucoma occurred in only the eyes with a cataract and occurred in both eyes in bilateral cataracts unless otherwise stated. All the children had surgery apart from the child with bilateral anterior segment dysgenesis. All types of glaucoma were detected postoperatively apart from the child with Lowe's syndrome and the child with bilateral ocular hypertension..

Table 31: The number of children with different types of glaucoma

	Bilateral cataract	Unilateral cataract
Postoperative Open Angle	16[α]	3
Postoperative Closed Angle	1[β]	3
Anterior Segment Dysgenesis	1	1
Lowes syndrome	1	
Ocular Hypertension	1	1
Retained haelon plugging angle	1[χ]	
Not Known		1
Total	21	9

[α] 7 children with unilateral glaucoma; 8 with bilateral glaucoma; 1 child with postoperative closed angle in one eye and open angle in the other

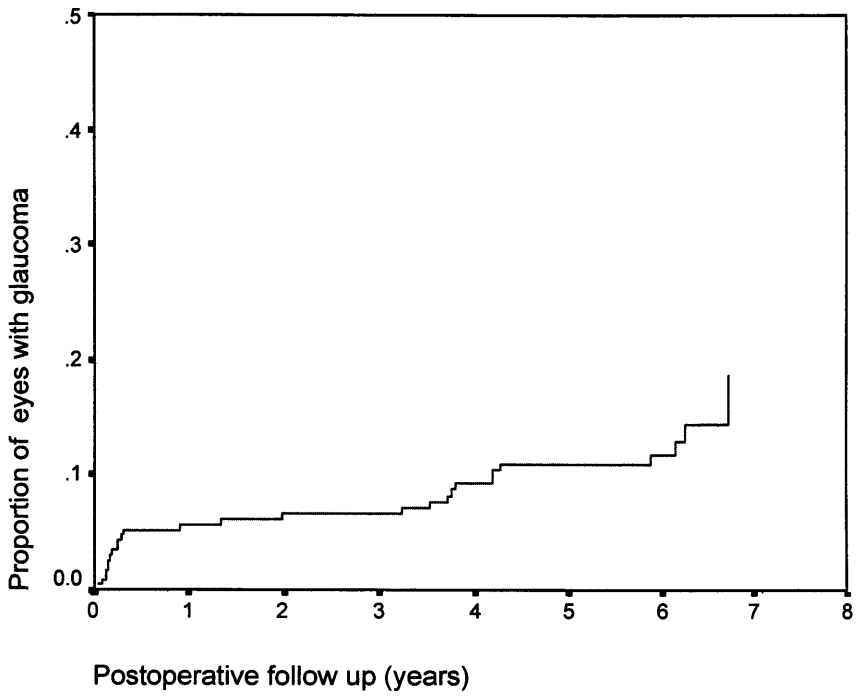
[β] 1 child with unilateral glaucoma
[χ] child with raised pressure in one eye only

5.3.2.1.1 Postoperative open angle glaucoma

27 (24 bilateral, 3 unilateral) eyes of 19 children developed postoperative open angle glaucoma. The median time to development of postoperative glaucoma was 1.34 years, with a range of 0.39 months to 6.73 years.

The proportion of eyes developing glaucoma over time is shown in Figure 56. The incidence rate of postoperative glaucoma in this study was 5.25 cases per 100 eyes undergoing cataract extraction per year.

Figure 56: Nelson-Aalen (Hazard) curve showing the development of postoperative glaucoma over time



A priori, the relationships between age at detection, age at cataract surgery, cataract surgery procedure, vitrectomy at primary procedure, primary intraocular lens implantation, significant postoperative uveitis, microphthalmia, severity of cataract at presentation, gender and laterality were investigated (Table 32).

Table 32: Association between the factors of interest in relation to postoperative glaucoma

	age at detection	age at surgery	cataract surgery	primary vitrectomy	primary IOL	postop uveitis	microphthalmia	severe cataract	gender	laterality
age at detection		0.78α	37.25β	-5.34γ	-2.27 γ	-2.18 γ	-2.53 γ	-2.00 γ	-0.19 γ	-0.11 γ
		p=0.00	p=0.00	p=0.00	p=0.02	p=0.03	p=0.01	p=0.05	p=0.85	p=0.92
age at surgery			69.92β	-5.84γ	-10.92γ	-2.94γ	-2.83γ	-5.66γ	-0.58γ	-0.76γ
			p=0.00	p=0.00	p=0.00	p=0.00	p=0.01	p=0.00	p=0.56	p=0.45
cataract surgery				261.49δ	74.78δ	0.80δ	2.84δ	12.07δ	4.06δ	0.08δ
				p=0.00	p=0.00	p=0.67	p=0.24	p=0.00	p=0.13	p=0.96
primary vity					48.34δ	0.58δ	0.38δ	3.46δ	3.22δ	0.01δ
					p=0.00	p=0.45	p=0.54	p=0.06	p=0.07	p=0.92
primary IOL						7.69δ	4.00δ	28.95δ	0.44δ	0.02δ
						p=0.06	p=0.05	p=0.00	p=0.51	p=0.88
postop uveitis							1.23δ	5.06δ	1.82δ	0.21δ
							p=0.27	p=0.03	p=0.18	p=0.65
microphthalmia								4.31δ	4.51δ	5.86δ
								p=0.04	p=0.04	p=0.02
severe cataract								2.45δ	0.13δ	1.76δ
									p=0.12	p=0.72
gender										1.23δ
										p=0.27

These variables were factors that could possibly influence postoperative open angle glaucoma.

There was a significant positive correlation between age at detection and age at surgery(0.78, p=0.00), as earlier detection would allow earlier surgery. Age of detection and age of surgery were also highly associated with surgical techniques(type of surgery (37.25 p=0.00; 69.92, p=0.00), primary vitrectomy (-5.34, p=0.00;-5.84, p=0.00) and IOL implantation(-2.27, p=0.02;-10.92, p=0.00)). This may be because the age of an infant may determine the type of surgery, e.g. a very young infant probably would not have an IOL implantation, and lensectomy/vitrectomy would therefore be appropriate.

Age at detection and age at cataract surgery were also highly associated with microphthalmia(-2.53, p=0.01; -2.83, p=0.01) and visually significant cataract (-2.00, p=0.05; -5.66, p=0.00), perhaps reflecting the ability to pick up more obvious abnormalities by the clinicians and the earlier opportunity to conduct surgery. Postoperative uveitis was associated with age at detection(-2.18, p=0.03) and age at surgery(-2.94, p=0.00) as well as severe cataract at detection(5.06, p=0.03). This may reflect a greater uveitic response postoperatively in younger children.

Univariately, the distribution of cases of postoperative open angle glaucoma and the above factors are shown in Figures 57-64, Appendix 7.

As described in the Methods section (*refer to section 4.3.3.1.2 Glaucoma, page 85*), a Cox regression model (Table 33) was used to investigate the association between postoperative glaucoma with age at detection, age at cataract surgery, cataract surgery procedure, vitrectomy at primary procedure, primary intraocular lens implantation, significant postoperative uveitis, microphthalmia, visually significant cataract, gender and laterality. Of these, microphthalmia and primary intraocular lens implantation, together with age at surgery and age at detection were significant or approached significance in univariate analysis at the 0.05 level and are highlighted in bold.

Table 33: Cox regression univariate analysis showing the association between time to glaucoma and variables of interest

Variable	Hazard Ratio (95% confidence interval)	P value	Clustering within child 95% CI (p value)
Log(Age at detection in days)	**0.36 (0.21 to 0.59)**	**0.000**	**0.20 to0.59 (0.00)**
Log(Cataract surgery age in days)	**0.31(0.16 to 0.58)**	**0.000**	**0.15 to 0.63 (0.00)**
Type of cataract surgery (Lens aspiration alone)# (baseline)			
Type of cataract surgery (Lens aspiration and vitrectomy)#	0.45(0.17 to 1.22)	0.12	0.14 to0.1.48 (0.19)
Type of cataract surgery (Lensectomy-vitrectomy)#	1.60(0.53 to 4.61)	0.42	0.50 to 4.63 (0.48)
Primary vitrectomy	2.04(0.77 to 5.40)	0.150	0.65 to 6.47 (0.22)
Primary intraocular lens implantation	**0.36(0.12 to 1.04)**	**0.06**	**0.10 to 1.31 (0.12)**
Significant postoperative uveitis	1.17(0.16 to 8.70)	0.88	0.14 to 8.75 (0.92)
Severe cataract at detection	22.92(0.032 to 16618.30)	0.35	0.021 to 16632.51 (0.37)
Microphthalmia	**2.47(1.05 to 5.85)**	**0.04**	**0.82 to7.45 (0.11)**
Gender	1.53(0.71 to 3.30)	0.28	0.60 to 3.92
Laterality	0.65(0.19 to 2.14)	0.47	0.19 to 2.24 (0.49)

#Lensectomy-vitrectomy vs Lens aspiration alone (Hazard ratio= 0.71CI=0.19 to 2.63, p=0.60; intereye correlations CI=0.14 to 2.71, p=0.61)

In the multivariate model however, age at detection remained the only independently associated factor. A 10-fold increase in the age at detection (for example, 10 days compared to 1 day or 60 days compared to 6 days) was associated with a 64% decrease in the hazard ratio (95% CI=41%-79%, p<0.001). However, 3 of the variables that are not significant in the multivariate model (age at cataract surgery, p=0.42; microphthalmia, p=0.17 and primary IOL, p=0.81,) are highly correlated with age at detection as shown in Table 32 previously (Spearman's correlation coefficient=0.78, p=0.00; Z=-2.53, p=0.01; Z=-2.27, p=0.02 respectively). Furthermore the hazard ratio univariately for microphthalmia (2.47(1.05 to 5.85)) and primary IOL (0.36(0.12 to 1.04)) are high and so these are probably still clinically significant factors. Thus the final multivariate model retained these factors and is shown in Table 34.

Table 34: Multivariate cox regression model of variables associated with development of glaucoma

Variable	Hazard ratio (95% confidence interval)	P value	Clustering within child 95% CI (p value)
Log (Age at detection)	0.36 (0.21 to 0.60)	0.000	0.22 to 0.59 (0.00)
Log (Cataract surgery age)	0.70 (0.30 to1.65)	0.42	0.21 to 2.30 (0.56)
Microphthalmia	1.84 (0.77 to 4.39)	0.17	0.55 to 6.20 (0.32)
Primary intraocular lens implantation	1.17 (0.33 to 4.11)	0.81	0.21 to 6.64 (0.86)

Therefore the findings of this study based on this model suggest, that after taking into account the age of detection, if a child of 1 week of age waited until they were 1 month old to have surgery, it would decrease their hazard ratio by 37%. If the child had microphthalmia it would increase the hazard ratio by 184% of that without microphthalmia. A primary IOL increases the hazard ratio to a lesser extent (117%) of that without a primary IOL implantation which contradicts the findings of the univariate analysis in which a primary IOL implantation decreased the hazard ratio (0.36).

The following Nelson-Aalen (Hazard) curves (Figures 65-68) illustrate how the risk of postoperative glaucoma alters if these significant variables are taken into account. The graphs were truncated at postoperative follow up of 6.95 years.

Figure 65: Nelson-Aalen (Hazard curve) showing the development of glaucoma over time by age at detection

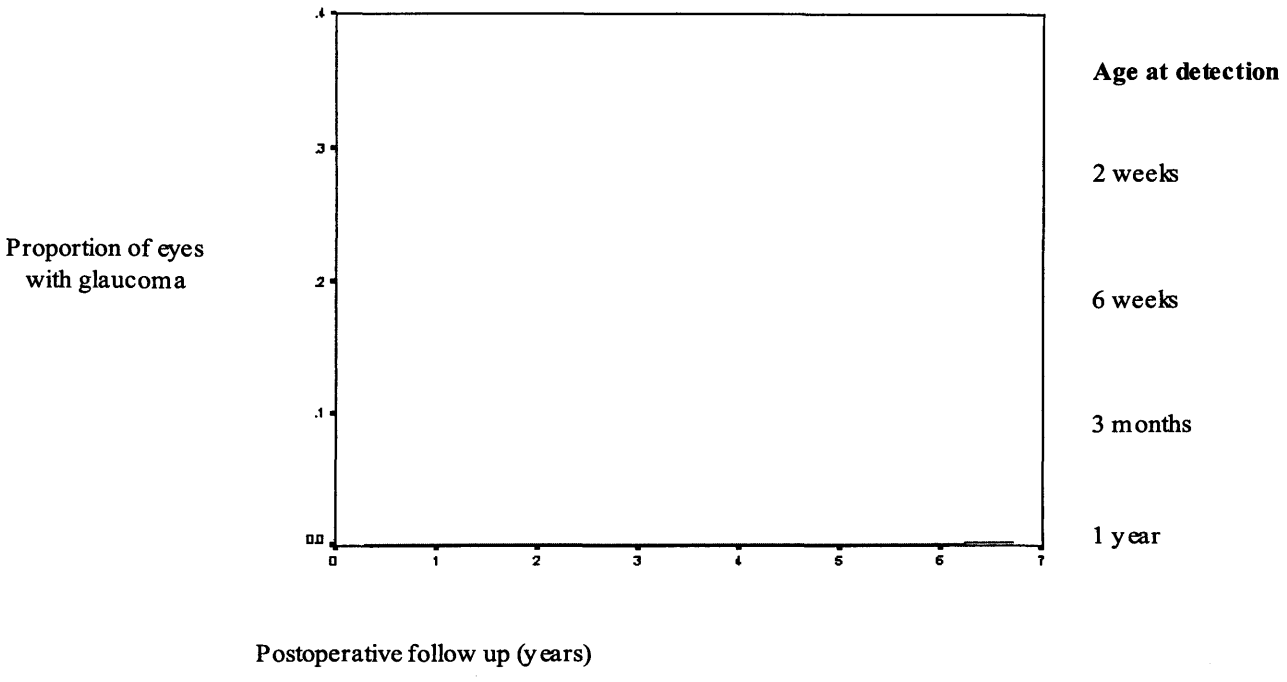


Figure 66: Nelson-Aalen (Hazard curve) showing the development of glaucoma over time by cataract surgery age

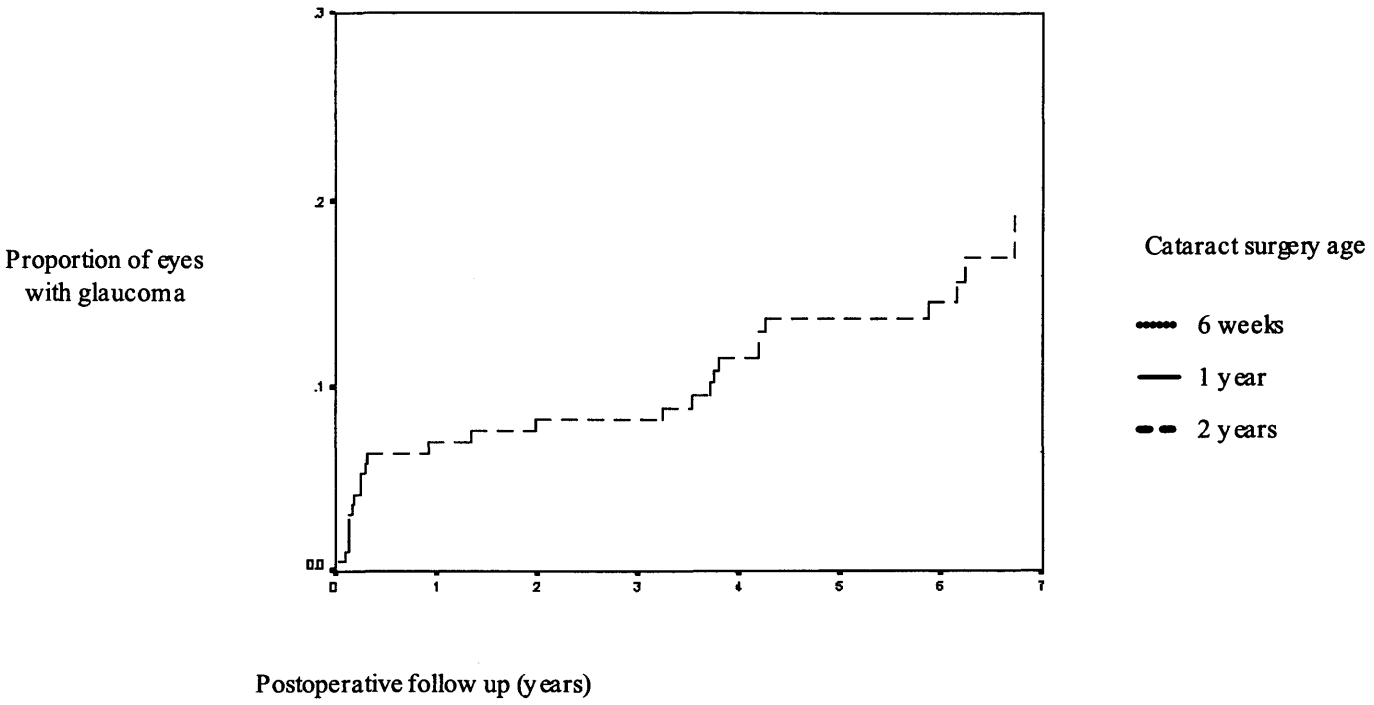


Figure 67: Nelson-Aalen (Hazard) curve showing the development of postoperative glaucoma over time for a child undergoing cataract surgery at 6 weeks according to presence of microphthalmia

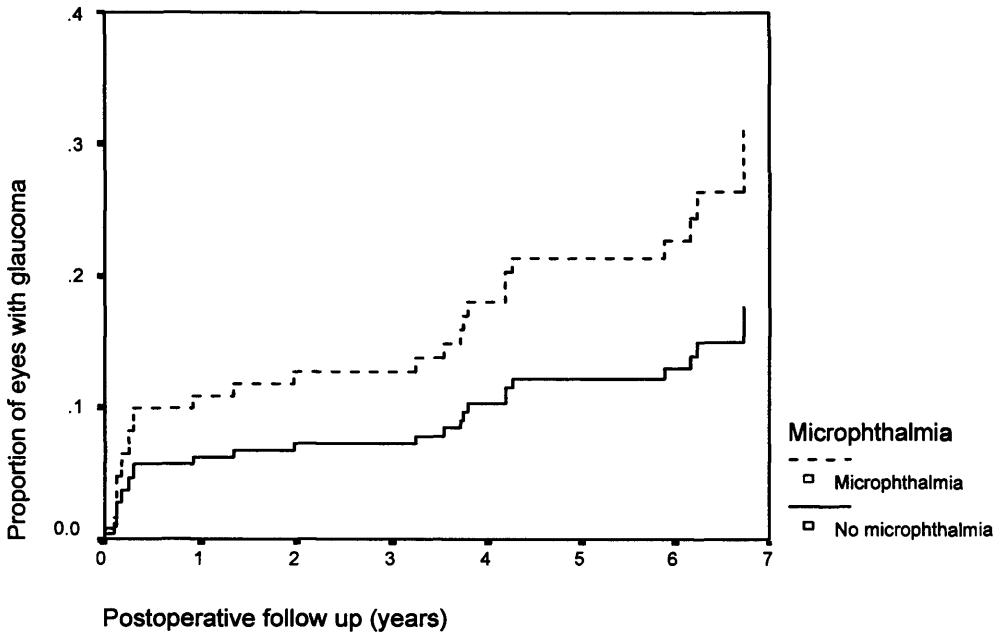
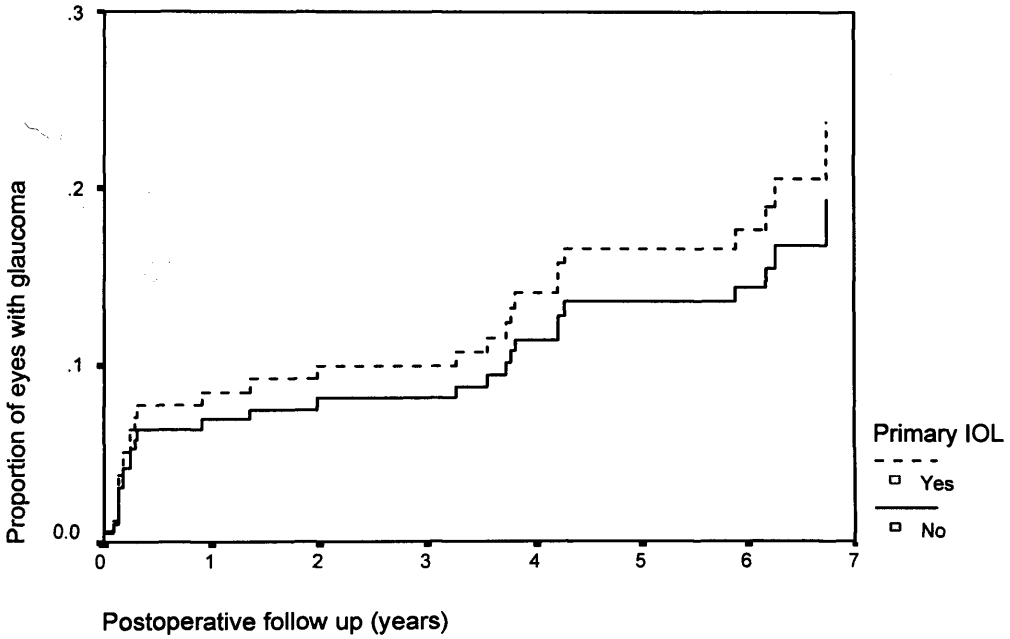


Figure 68: Nelson-Aalen (Hazard) curve showing the development of postoperative glaucoma over time for a child undergoing cataract surgery at 6 weeks according to presence of primary intraocular lens implantation



5.3.2.2 Other postoperative complications

One child (girl) with bilateral cataracts developed endophthalmitis. Cataract surgery (lens aspiration, no vitrectomy and primary IOL) was performed at 8.44 years. VA at follow up was perception of light (PL).

4 eyes of 3 children had a retinal detachment. All were operated on at less than one year of age and all had a vitrectomy. The final visual acuity of all these eyes was very poor (Table 35).

Table 35: Number of children with postoperative retinal detachment

Gender	Aetiology of cataract	Cataract surgical procedure	Method of posterior capsulecto-my	Primary IOL implant	Age of cataract surgery	Treatment of retinal detach-ment	Visual acuity
Boy	Trisomy 21	Lensectomy/ vitrectomy	Mechanical	No	8 days	None	NPL
Boy	Trisomy 21	Lensectomy/ vitrectomy	Mechanical	No	8 days	Vitrectomy	NPL
Girl	Galactosae-mia carrier	Lens aspiration and vitrectomy	Mechanical	No	81 days	Vitrectomy	HM
Boy	Idiopathic	Lensectomy/ vitrectomy	Not known	No	49 days	Vitrectomy	NPL

As shown in Table 36, 9 children with bilateral cataract had excessive postoperative inflammation, defined as that which required more medication than the normal postoperative regime. In all cases this involved one eye and the child had no associated risk factors for increased inflammation such as any systemic diseases associated with uveitis. They were operated on late apart from one child at 4 months and had a range of surgical procedures, and 67% had an intraocular lens implantation. The small number of cases precluded further analysis to investigate possible risk factors.

Table 36: Number of children with postoperative uveitis

Gender	Aetiology of cataract	Cataract surgical procedure	Primary IOL implant	Age of cataract surgery	Normal regime	Procedure to treat uveitis	Visual acuity at last followup
Girl	Isolated	Lensectomy/vitrectomy	Yes	6.35 years	Maxidex & cyclopentolate QDS	Increased topical meds	6/48
Boy	Isolated	Lens aspiration and vitrectomy	Yes	5.29 years	Predforte & cyclopentolate	Increased topical meds	6/6
Boy	Isolated	Lens aspiration alone	Yes	5.27 years	Predforte & cyclopentolate	Increased topical meds and floor and subconj injections	6/6
Boy	Isolated	Lensectomy/vitrectomy	No	4 months	Betnesol & phenylephrine	Increased topical meds	PL
Girl	Ocular	Lensectomy/vitrectomy	No	1.04 years	Predforte in a reducing dose	Increased topical meds	6/18
Girl	Isolated	Lens aspiration and vitrectomy	Yes	7.86 years	Maxitrol QDS in a reducing dose	Increased topical meds	6/6
Girl	Isolated	Lens aspiration alone	Yes	5.54 years	Maxidex & betnesol	Increased topical meds and subconj injection	6/9
Girl	Isolated	Lens aspiration alone	Yes	3.72 years	Predforte in a reducing dose	Increased topical meds	6/36
Boy	Isolated	Lens aspiration alone	Yes	3.81 years	Not known	Increased topical meds	6/9

The numbers of children developing posterior capsular opacity or secondary membrane are shown in Table 37. 27 children were boys and 20 were girls. Median age of cataract surgery was 285 years (10 days to 11.36 years). Median visual acuity at follow up examination was better for children with PCO 6/9 (6/5 to PL) than those developing secondary membranes 6/36 (6/9 to PL).

Table 37: Children developing posterior capsular opacity or secondary membrane (N=47)

	Bilateral cataract	**Unilateral cataract**
Posterior capsular opacity	35 children* 54 eyes(23%)	7 children 7 eyes(18%)
Secondary membrane	4 children* 5 eyes(2%)	3 children 3 eyes(7%)

*2 children had PCO in one eye and secondary membrane formation in the other

5.3.3 Quality of life of children with congenital cataract

41/42 parent-child pairs invited to participate, completed questionnaires (33 were parental-child complete pairs and 8 were parental completion only). 14 families completed the questionnaires in clinic and 28 completed them at home. The demographics of the children are outlined above *(refer to section 5.2.2.1.Parental perception of the effects of amblyopia treatment on their children, page 112)* .

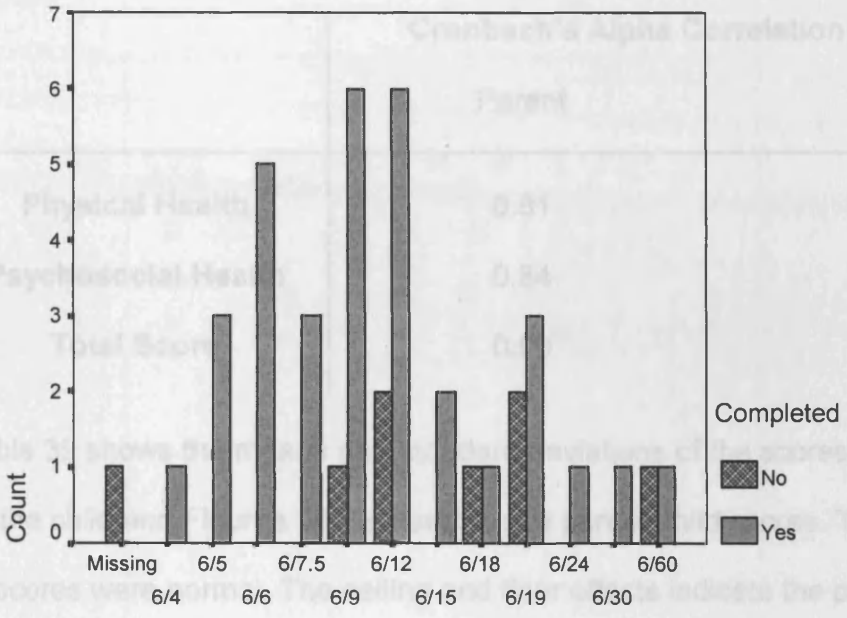
5.3.3.1 The PedsQL 4.0™

To assess the feasibility and practicality of administration of the PedsQL 4.0™ Generic Core Scales, the percentage of missing values was calculated. Of completed forms, the percentages of missing item responses was very low, being 0.66% for children and 0.95% for their parents.

8 children failed to complete any part of the QOL forms. These children tended to have poorer visual acuity (Figure 69) and 3 had Down's syndrome, 1 had dental anomalies, 1 had a systolic murmur and the remaining 3 had no other medical conditions.

One child with reduced vision (best VA=6/12) commented on the yellow paper of the forms being too bright, making the words difficult to read. Otherwise there were no comments on the questionnaires themselves and the ease of completion.

Figure 69: Comparison of the distribution of best visual acuity of children who failed to complete the questionnaire(N=8) with those that did (N=33).



Internal consistency reliability alpha coefficients for the PedsQL 4.0™ exceeded the minimum reliability standard of 0.70 for group comparisons[202] and exceeded or approached the minimum reliability standard of 0.90 for individual comparisons[202] as shown in Table 38.

Table 38: Cronbach's Alpha Correlation Coefficient for physical, psychosocial and physical health

	Cronbach's Alpha Correlation Coefficient	
	Parent	Child
Physical Health	0.81	0.88
Psychosocial Health	0.84	0.90
Total Score	0.90	0.93

Table 39 shows the means and standard deviations of the scores by proxy and by the child and Figures 70-72 illustrate the parent-child scores. The distribution of scores were normal. The ceiling and floor effects indicate the proportion of children scoring the maximum (100) and minimum (0) possible scores and they indicate the potential difficulties in interpreting results when scores of the study group are nearly perfect (ceiling) or very poor (floor).

Table 39: Mean, median, ceiling and floor effects of the Parental and child PedsQL 4.0TM scores

	Mean (SD)		Median		% Floor Effects		% Ceiling Effects	
	Parent	Child	Parent	Child	Parent	Child	Parent	Child
Physical Health	80.22 (22.01)	80.76 (18.61)	87.50	87.50	0	0	24.39	5.00
Psycho-social Health	73.52 (16.44)	72.93 (16.06)	73.33	72.93	0	0	0	0
Total Score	75.91 (16.79)	75.85 (15.56)	79.35	75.85	0	0	0	0

Figure 70: Distribution of child and parent pairs physical health summary scores

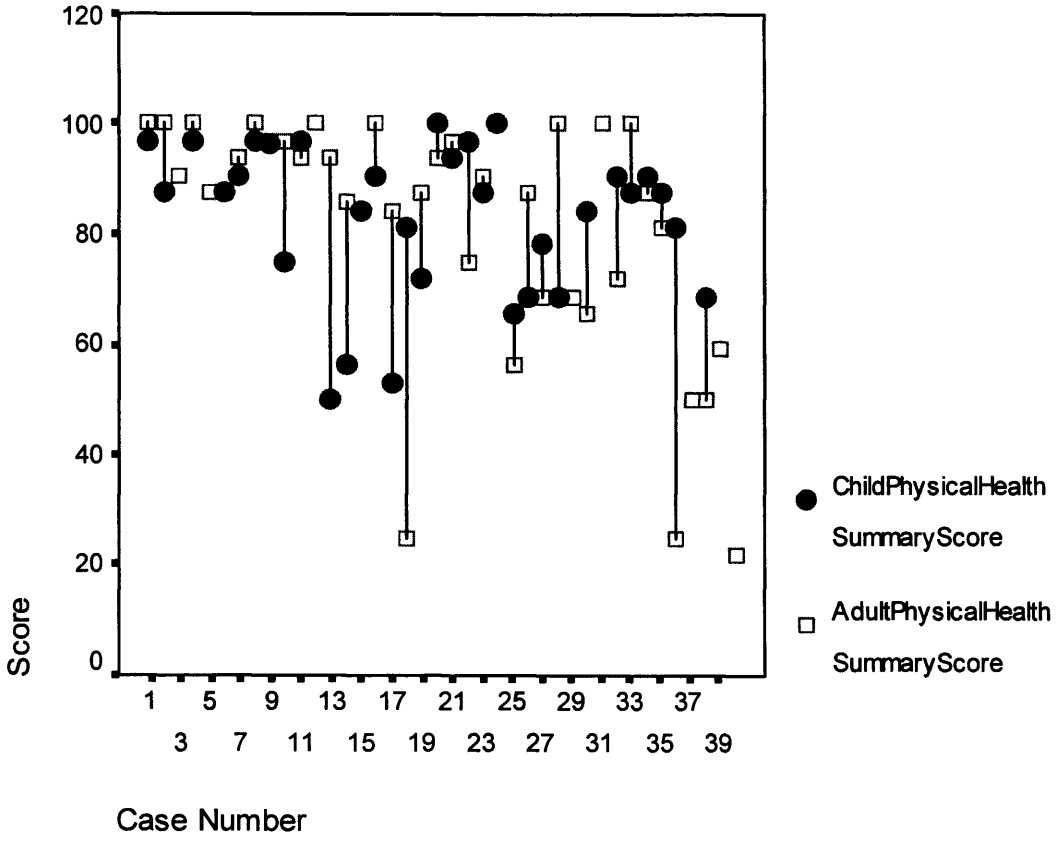


Figure 71: Distribution of child and parent pairs' psychosocial health summary scores

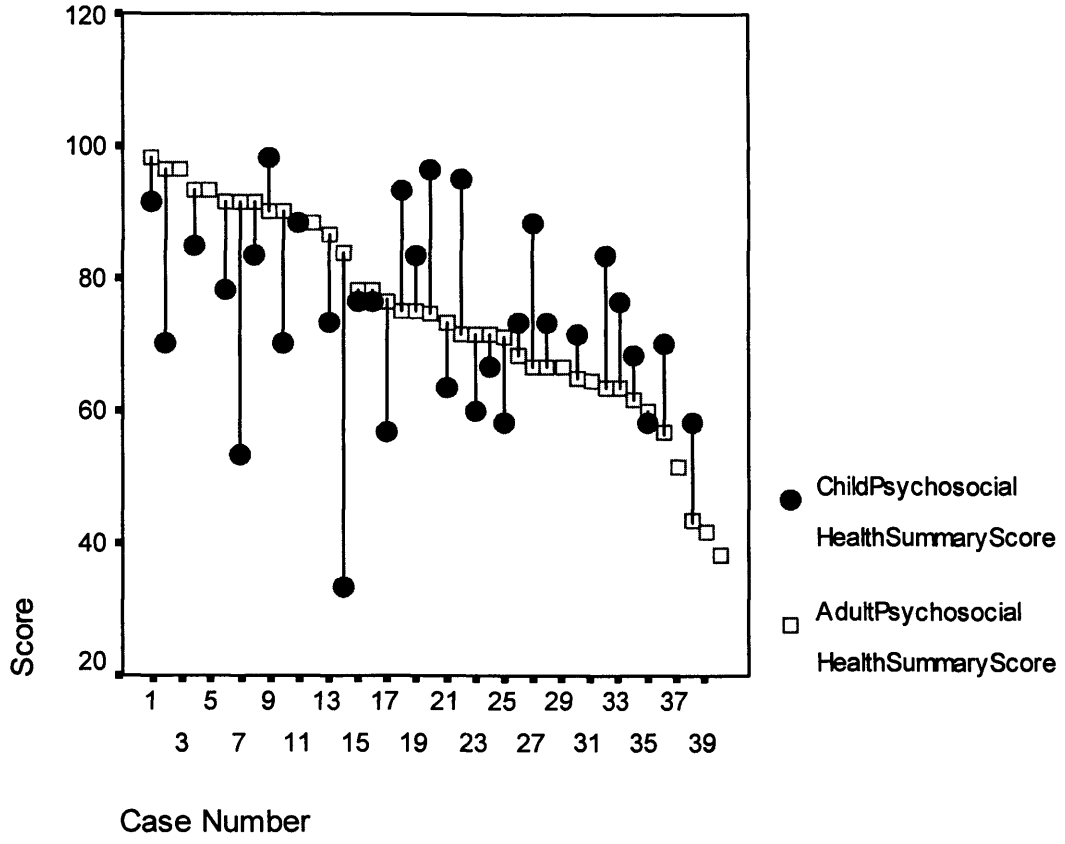
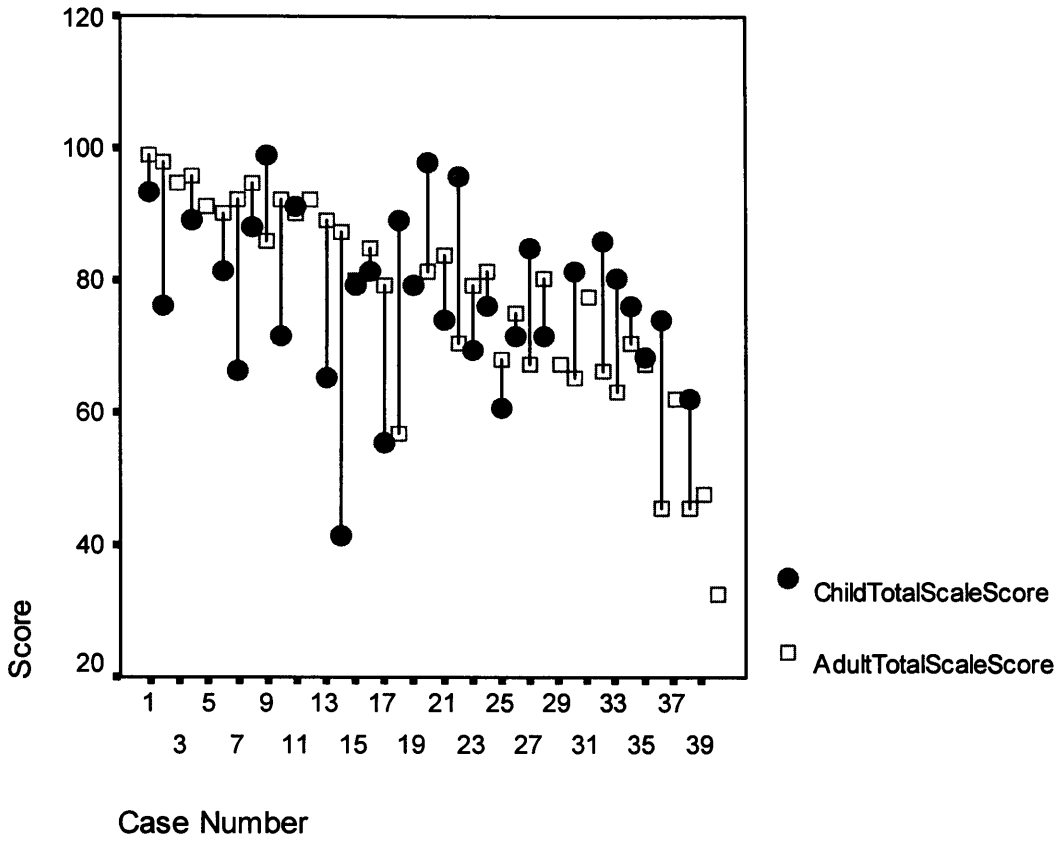


Figure 72: Distribution of child and parent pairs' total scale scores



No association was found between the physical, psychosocial or total summary mean scores respectively and age, sex, laterality of cataract, best visual acuity achieved (uniocular or binocular), presence/absence of other medical disorders, surgery, strabismus, significant postoperative complications, glaucoma, posterior capsular opacity or amblyopia treatment. However the sample may have been too small to discern any true association.

Child and parental agreement was assessed using the Bland Altman[203] measure of agreement Table 40. Although the mean difference between the parent and child was quite small (up to –2.0) the range of disagreement was wide.

Table 40: Bland-Altman measure[203] of agreement of child-parent pair scores of physical, psychosocial and total scale scores

	Min	Max	Mean	Lower limit of agreement	Upper limit of agreement
Difference in physical score (child-parent)	-43.8	56.3	-1.6	-44.7 CI=-31.2 to-58.3	41.6 CI=28.0 to 55.1
Difference in psychosocial score (child-parent)	-50.6	23.3	-2.0	-35.8 CI=-25.2 to-46.4	31.8 CI=21.2 to 42.4
Difference in total score (child-parent)	-46.2	32.2	-1.3	-36.7 CI=-25.8 to-47.6	34.1 CI=23.2 to 45.0

There was no relationship between the mean score of a child and their parent and the difference between the pair (child-adult). Furthermore, Figures 73-75 illustrate the degree of lack of agreement (up to 56.3 points difference) between some children and their parents for physical, psychosocial and total scale scores.

Figure 73: Physical summary scores of child and parent pairs

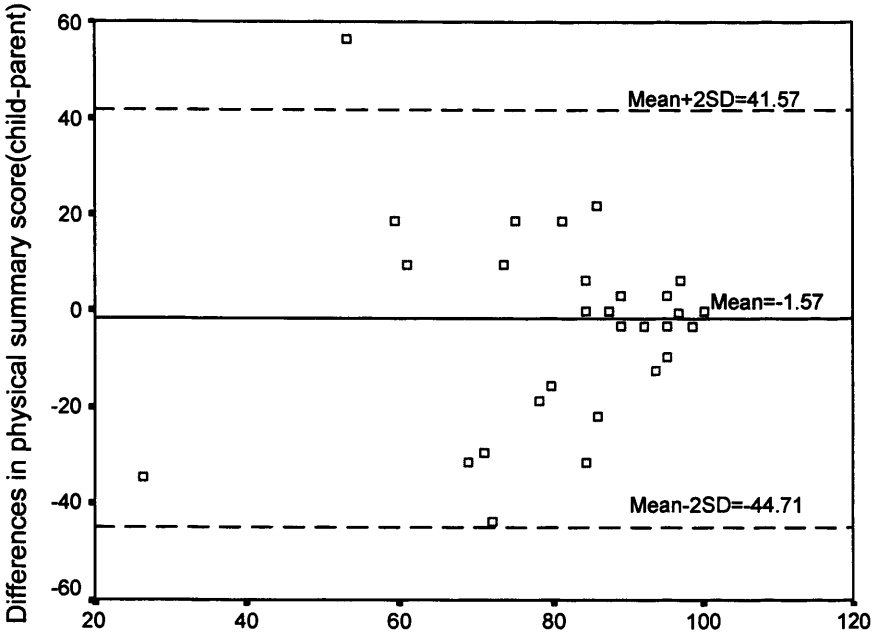


Figure 74: Psychosocial summary scores of child and parent pairs

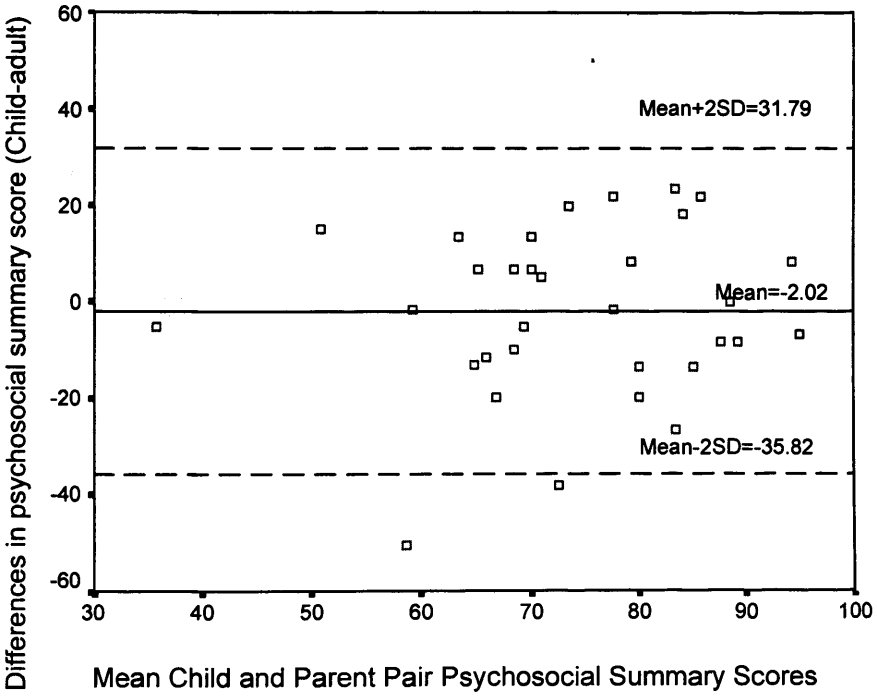
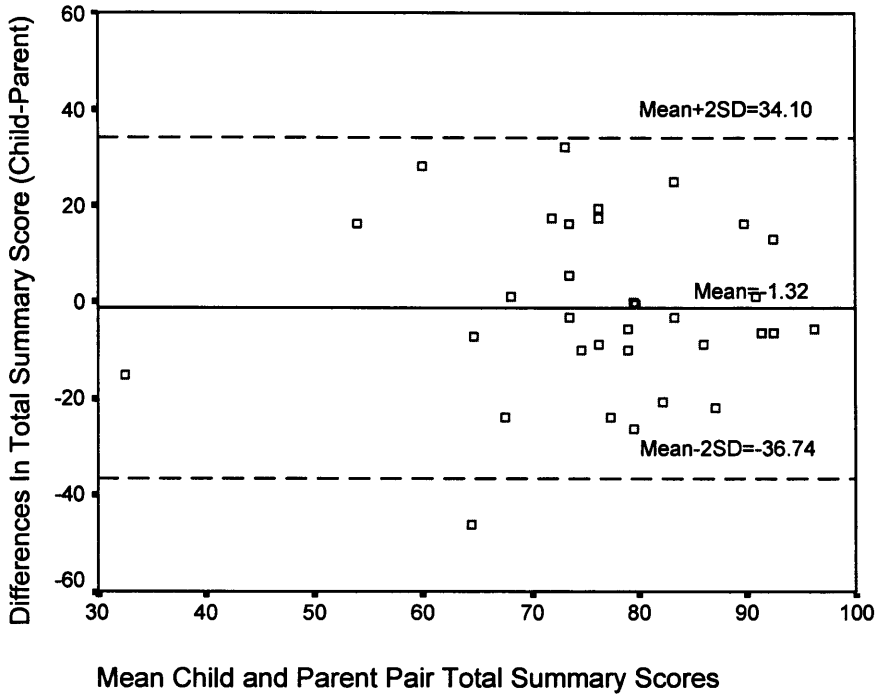


Figure 75: Total summary scores of child and parent pairs



The psychosocial PedsQL scores of children with congenital cataract in the present study were comparable to those reported by children with notably more debilitating or life threatening diseases such as childhood cancers[224] and rheumatological disease[225] as shown in Table 41. For example, the scores of the children with congenital cataract and their parents were consistently lower in all modalities compared to children with acute lymphoblastic leukaemia[224].

Table 41: PedsQL 4.0™ child scores (total, physical and psychosocial) of children with congenital cataract, rheumatological disorders and paediatric cancers

	Mean (SD) Physical Health	Mean (SD) Psychosocial Health	Mean (SD) Total Summary Score

Congenital cataract	80.76 (18.61)	72.93 (16.06)	75.85 (15.56)
Rheumatological disorders[225*]	68.12(22.52)	74.23(16.46)	72.09(16.92)
Paediatric cancers[226+]	71.79(21.80)	72.62(16.41)	72.20(16.38)
Acute lymphoblastic leukaemia[224]	86.16(14.72)	79.07(13.14)	80.97(12.57)

Table 42: PedsQL 4.0™ parental scores (total, physical and psychosocial) of children with congenital cataract, rheumatological disorders and paediatric cancers

	Mean (SD) Physical Health	**Mean (SD) Psychosocial Health**	**Mean (SD) Total Summary Score**
Congenital cataract	80.22 (22.01)	73.52 (16.44)	75.91 (16.79)
Rheumatological disorders[225*]	70.97(18.49)	66.72(24.12)	73.31(17.62)
Paediatric cancers[226+]	68.75(24.98)	70.31(17.96)	69.70(19.17)
Acute lymphoblastic leukaemia[224]	84.73(19.73)	73.71(16.95)	76.50(16.10)

*dermatomyositis, fibromyalgia, juvenile rheumatoid arthritis, systemic lupus erythematosus, spondylarthritis
+acute lymphocytic leukaemia, brain tumour, non-Hodgkin's lymphoma, Hodgkin's lymphoma, Wilms' tumour and other cancers

6 DISCUSSION

6.1 Visual Acuity Outcomes

In this study, as expected, the best visual acuities at follow up were of non-cataractous eyes of children with unilateral cataract as there are no amblyogenic factors. The visual acuities of cataractous eyes of children with unilateral cataract however, had the worse visual outcome despite surgery (median=6/48, range=6/5 to NPL) as these eyes had both visual deprivation and competitive amblyopia. Visual acuity of cataractous eyes of children with unilateral cataracts managed conservatively, (median=6/24, range=6/5 to PL) were better than those with intervention. This is probably a reflection of less severe cataracts in the non-surgical group (Chi-squared=7.74, p=0.005). These findings suggest that clinicians should be guarded about visual prognosis to parents of children with unilateral cataract. Poor visual outcomes despite surgery and aggressive amblyopia treatment may not in fact provide the useful "spare" eye that clinicians often propose as the main reason for treating unilateral cases.

Children with bilateral cataract who underwent surgery had a better visual acuity by eye than those with unilateral cataracts (median=6/19, range=6/5 to PL) as there was no competitive amblyopia between the eyes. As with the children with unilateral cataracts, eyes of children with bilateral cataracts that were managed conservatively had a better visual acuity (median=6/12, range=6/4 to NPL) than those undergoing surgery, reflecting less severe cataract in this group (Chi-

squared=136.91, p=0.00). The majority of children with bilateral cataract in this study would achieve a visual acuity by eye of at least 6/18. 6/18 or better is the visual acuity at which children can be expected to be educated at a normal school with minimum extra help[59]. This finding will help inform future parents of children with congenital cataract and has implications for resource allocation in terms of schooling and additional care.

The reliability of these findings is dependent on the children being examined, the examiner's methods as well as the consistency and dependability of the method used to measure the visual acuity. The reliability of visual acuity recorded in children with reduced vision appears to be as reliable as the reliability in adults (mean age=4.8 years)[53;56]. The reliability also appears to improve with age as children become more literate, mature and familiar with the visual testing protocols and this is also true of amblyopic eyes[204]. To take account of this issue in this study, young children below the age of 5 were excluded from analyses relating to visual acuity. Although a large number of examiners and units were involved in the study, the vast majority were examined in specialist units with expert assessment of visual acuity. Thus the results therefore reflect prevailing practice and as such can be viewed pragmatically as functional outcomes which have been analysed appropriately.

This study describes the management of children in the UK nationally and direct comparisons with non-population based studies is difficult. Nevertheless, the visual acuity range of children with bilateral cataracts in this study were similar to those reported previously [59;154;205;206]. Although poor, the visual acuity of eyes with cataract of children with unilateral cataract is generally better than described in the literature[59;67;207]. This difference may reflect that many previous studies, based on small selected case series, were conducted over 15 years ago and that the outcomes of congenital cataract are improving with better surgical techniques and amblyopia treatment regimes. Support of a secular trend in outcomes comes from repeated studies of the causes of visual impairment in children attending a blind school in Scotland. In the survey in 2002 congenital cataract was not found, but accounted for 12/99 and 4/93 respectively in the 1980s and 1990s[208].

6.1.1 Factors associated with the visual acuity of children with bilateral cataract.

Visual acuity of children with bilateral cataracts was associated with a number of factors in the multivariate model *(refer to Table 25: Multivariate ordinal regression model of factors associated with visual acuity at follow up examination of 182 eyes of 98 children with bilateral cataracts).*

Most importantly, earlier surgery predicts better visual acuity in children with bilateral cataracts. Previous reports in the literature also recommend early surgery at different age points, including <6 weeks [205], <2 months[59], <8 weeks[166]

and <10 months[44]. Postoperative complications were also associated with a poorer visual outcome, as would be expected, however there was a significant association between age at surgery and postoperative complications, echoing the findings of other studies[59;205;206]. Timing of surgery of visually significant cataract must be within the critical period to prevent amblyopia *(refer to section 2.1.2 Visual development, page 21)*, however complications have been shown to increase in early surgery[93]. There is need therefore to balance the timing of surgery to prevent amblyopia with the best time to minimise postoperative complications. To determine this equilibrium, a prospective trial randomising children into different surgical age groups within the critical period is necessary to establish precisely the optimum time for surgery.

A poorer visual acuity was also associated with the presence of other medical conditions, which has also been previously reported[154;205]. This may be because vision is limited by learning disabilities or cortical impairment, rather than congenital cataract per se. Visual acuity assessment of such children will be difficult and postoperative care such as occlusion will also be challenging. In addition, repeat hospital appointments and inpatient care for other non-ophthalmic conditions, would disrupt postoperative treatment such as occlusion. However, some functional improvement is likely with treatment, even if it cannot be measured objectively, thus the presence of another medical condition should not preclude these children from the same care as children without medical conditions.

Better concordance with the occlusion regime was not associated with a better visual acuity univariately, but became significant in the multivariate model once the age of cataract surgery had been taken into account, as concordance with occlusion was statistically correlated with age at cataract surgery. Concordance with occlusion regimes is known to be an important factor in preventing amblyopia and factors to improve concordance are discussed below *(refer to section 6.3 Impact of Amblyopia Treatment, page 191).*

The presence of a more severe cataract was associated with a poor visual acuity in the univariate analysis (p=0.07). Multivariately, however, the presence of a severe cataract was associated with better visual acuity. This anomalous finding reflects the statistical correlation between age at cataract surgery and severity of cataract *(refer to Table 23: Association between the factors of interest in relation to visual acuity of children aged >5 years old with bilateral cataracts).* A more severe cataract will probably be more noticeable clinically, be detected early and be operated on early. Thus early cataract surgery is associated with better visual acuity as described above, and therefore, more severe cataract may be too. Indeed, one study suggested that the morphology of cataracts was the best predictor of visual outcome and that other possible associated factors were actually constituent features of cataract type[14].

6.1.2 Factors associated with the visual acuity of children with unilateral cataract.

The two models of factors associated with visual acuity in children with bilateral and children with unilateral cataracts differ reflecting the different nature of the disease in terms of development of amblyopia *(refer to section 2.2.2 Management of congenital cataract by the laterality of the cataract, page 34).* Although both models contained concordance with occlusion treatment as a factor significantly associated with visual acuity, in the unilateral cataract model this was the only statistically significant factor and the hazard ratio was higher than in the bilateral cataract model (7.92 compared to 5.64). This supports the suggestion previously reported in the literature that concordance with amblyopia treatment is perhaps the most important predictor of vision[8;154;155] *(refer to section 2.2.5.2 Occlusion and penalisation, page 47).* It is recognised that in children with unilateral cataract, occlusion is difficult as the amblyopia is so dense: in one series, 21/30 children abandoned the regime[207]. It is therefore important to help determine the reasons for poor concordance and this issue is discussed further below *(refer to section 6.3 Impact of Amblyopia Treatment, page 191).*

In this study, age at detection was not statistically significant in the multivariate model. However, age at detection was correlated with the age at surgery, type of cataract surgery, aetiology of the cataract, severity of the cataract and postoperative complications. It may be the interactions of all of these factors with occlusion concordance that resulted in it not being significant in the final model as univariately, an earlier age at detection was associated with a better visual

acuity. It is known physiologically and from clinical experience that to prevent visual deprivation amblyopia, the cataract should be removed within a critical period *(refer to section 2.1.2 Visual development, page 21)*. Early detection is therefore crucial to allow timely intervention to occur[59] and the importance of continuing with effective screening of babies within this critical period remains high *(refer to section 2.1.4.2 Secondary strategies, page 28).* Less than half of this study cohort were detected at the routine newborn and 6-8 week examinations as reported previously[47]. Variations exist in the practices and training of paediatricians currently responsible for these examinations *(refer to section 2.1.4.2 Secondary strategies, page 28).* Implementation of training schemes and supervision for screening paediatricians are necessary. Audits of these schemes should be undertaken to evaluate their effectiveness. Routine monitoring of the screening process at a national level would ensure that standards are maintained.

Formal screening programmes could be augmented by parental input to improve detection of congenital cataract and might be particularly effective in familial cases. Parents often suspect an ophthalmic problem before a health professional becomes aware of it, more so in more cosmetically obvious presentations[209]. However, parental concern does not always ensure early diagnosis by health professionals[47]. The role of parents in the early detection of congenital cataract could be strengthened by public health campaigns to increase awareness and

knowledge of abnormal features. Increased awareness in health professionals to respond actively to these concerns should also be promoted.

The use of IOLs have been advocated especially in children with unilateral cataract in terms of providing a better refractive correction thereby reducing amblyopia and improving visual acuity[100;210] *(refer to section 2.2.5.1 Refractive Correction, page 43).* There was no association between visual acuity of children with bilateral or unilateral cataract and a primary intraocular lens implantation in this study. Furthermore, contact lenses were an acceptable and effective form of refractive correction in this study as reported by others[129]. Thus the findings of this study supports the continued practice of lensectomy/vitrectomy and contact lens correction until prospective work, preferably randomised control trials comparing this approach with lens aspiration and IOL show demonstrable positive benefits of the latter.

6.2 Postoperative open angle glaucoma

The annual incidence of postoperative open angle glaucoma in this study was 5.25% of eyes operated/year. Prevalence from other studies varies from 6 to 30%, depending on the series and length of follow up[85;86;88-90;93]. As the median follow up time of children that did not develop glaucoma was 6.05 years, there may have been an underestimation of the number of cases developing glaucoma as it can be as high as 46% at 13 years follow up[89].

Reports in the literature have used different criteria for the classification of postoperative open angle glaucoma. Some have used different levels of IOP[91;211] and some have classed a raised IOP alone as ocular hypertension[91]. This reflects the difficulty in diagnosis of children with glaucoma as examination of the discs and IOP assessment can be challenging and young children are unable to perform visual field tests. Inclusion of a case of postoperative open angle glaucoma into this study was based on treatment of glaucoma with topical medications and/or surgery and/or laser. This criterion was likely to define true cases as treatment was already initiated. This makes direct comparisons with other studies difficult. Furthermore, previous reports frequently do not take into account multivariate factors influencing each other[88-90] or have excluded one eye in the analysis of bilateral cases[91], and are largely based on selected case series only[212]. The present study therefore represents an arguably more accurate reflection of the incidence of glaucoma in the population, with appropriate multivariate analysis.

In this study, the median time to development of postoperative open angle glaucoma was 1.34 years (range=0.39 to 6.73 years). Importantly this is considerably less than the 2.6 to 12.2 year range reported from other studies[85;88-91;94;98;205;212-215]. It is already strongly advocated that children with congenital cataract should be followed up for life for signs of postoperative glaucoma. This study emphasizes that ophthalmologists should remain vigilant from the early post operative period.

The results of this study suggest that early age at detection is the most important factor associated with the development of glaucoma after congenital cataract surgery in this group of children. Previous studies have not addressed this factor specifically and therefore it is difficult to make direct comparisons. Age at detection is likely to be highly associated with other factors that have been well documented as possible risk factors in case series, and may in fact encompass many factors. The support for this comes from this study in which there was a significant association between age at detection and other factors univariately associated with postoperative open angle glaucoma: age at cataract surgery, microphthalmia and primary IOL insertion. In previous reports these factors have been identified as independent potential risk factors[14;86;89;90;97-99;212-214]. A small eye may be detected earlier and early surgery is only possible if the cataract is detected early on. Indeed in previous studies suggesting the most significant factor to be age at cataract surgery, the children had a disproportionately high proportion of other ocular abnormalities such as microphthalmia and also had

dense cataract[89]. Univariately, IOL implantation appeared to decrease the risk of glaucoma, however most children were implanted at older than 2 years of age *(refer to section 5.2.1.2 Intraocular lens (IOL) implantation, page 106).* In the and multivariate model when age at surgery was accounted for, IOL implantation increased the risk of glaucoma although this was not significant.

Thus paradoxically, the price of successful screening programmes to ensure early detection and treatment of congenital cataract within the critical period to prevent amblyopia *(refer to section 2.1.4.2 Secondary strategies, page 28)* may be an increased risk of postoperative glaucoma and other complications as surgery is performed earlier. However there must be an optimum period of time by which surgery can be delayed to reduce the risk of glaucoma and other postoperative complications without increasing the risk of amblyopia and further work is required to delineate this. Nevertheless the findings of the present study suggest that despite the adverse outcomes, surgery at an earlier age will result in better visual outcomes *(refer to sections 5.3.1.1 Factors affecting the visual acuity of children with bilateral cataracts, page 137 and 5.3.1.2 Factors affecting the visual acuity of children with unilateral cataracts page 145).* A retrospective study of 55 children with a mean follow up of 2.7+/-1.9 years suggested that the optimum time within the critical period for surgery resulting in the minimum amount of complications was less than 2 weeks of age[216]. A prospective trial of children randomised to cataract surgery in narrow age groups within the critical period would help to delineate precisely the optimum timing for surgery.

Despite contrary reports previously[85;86;211], there was no association in the present study between postoperative open angle glaucoma and the type of cataract surgery procedure. All three types of surgery were comparable with similar follow up periods, a previous criticism of other studies,[85;104]: aspiration and vitrectomy median=5.86 years(5 days to 7.34 years); aspiration alone, median=5.94 years (0.85 to 7.35 years); lensectomy/vitrectomy, median=6.19 (0.87 to 7.29 years). There was also no significant association between postoperative open angle glaucoma and postoperative uveitis or the severity of cataract at presentation as has been reported in some studies[14;86] but not by others[88;89;98].

6.3 Impact of Amblyopia Treatment

The psychosocial effects of amblyopia were investigated in 41 children. These children were recruited non-randomly from 6 centres around the country, which specialise in treating congenital cataract and are the major tertiary referral units for their regions in order to reflect the diversity of management in the UK. Nevertheless, there may be some selection bias, as the children who were included may have had additional complications or further surgery requiring specialist centre care rather than being referred back to their local hospitals for further follow up. The participation rate was high with the majority (41/42) questionnaires being fully completed and two thirds of the questionnaires were successfully completed at home which supports the use of the questionnaire in postal surveys as well.

Concordance with the occlusion regime was assessed through direct parental questioning and although validated by cross checking with the clinical notes it is likely that there may have been recall bias or an overestimation in the amount of occlusion achieved. A more reliable and objective measure of concordance would be achieved through the use of an occlusion dose monitor (ODM) (a modified occlusion patch connected to a data logger), as used in previous studies[217;218]. This would improve future investigations of the association between concordance and visual outcomes which is discussed below.

Although overall, contact lenses and glasses were comparable (61% positive reaction by the children in both groups), parents reported that glasses tended to have a negative effect on the child's interaction with their friends, which also affected the self confidence in some. This contrasted with reports from parents of children wearing contact lenses, who described them enjoying socialising whilst wearing the lenses and being more confident. These findings concur with other work suggesting that some older children do not wear glasses, despite an improvement in visual performance because of 'social' problems of wearing them[219] and some parents reported negative feelings towards their child wearing glasses[220].

Nevertheless, parents worried more about the contact lenses causing harm to their child than glasses. There are no directly comparable studies on this issue.

Effective refractive correction is essential to prevent amblyopia in children with congenital cataract. The impact of the different refractive correction methods on social relationships and parental worry about potential harm may have an impact on concordance with therapy. No association with concordance could be demonstrated in this study (*refer to Tables 18-22, page 129)*, but the size of the population studied may have limited the ability to detect a true effect.

In this study, 81% of the parents found occlusion difficult or very difficult and 32% thought their child had become more distressed or badly behaved as a result of

occlusion. Furthermore, a third of the parents thought that the relationship with their child had worsened as a result of occlusion. Despite this, the majority of parents (63%) never worried the patches were causing harm to their child. This concurs with a recent study of 364 parents which reported that occlusion was acceptable using a newly devised questionnaire (Amblyopia Treatment Index) which contained 3 subscales addressing attitudes to adverse effects of treatment, lack of treatment compliance and social stigma[167]. However another study using 2 instruments (Perceived Psychosocial Questionnaire and the Perceived Stress Index) contradicts these findings and found that the carer's stress and their child's psychosocial state were not influenced by occlusion[220]. None of the children in these two studies however had congenital cataract and it would therefore be of great interest to apply these new instruments to a population such as that in the present study.

In this study, no association was found between occlusion concordance and the child's or parental experience of occlusion as reported by the parents. This contrasts with other studies of children with amblyopia (not due to congenital cataract) which support the theory that parental perceptions of occlusion affect concordance and furthermore speculate that parental stress is a reflection of the child's distress[168]. It is postulated by others that concordance may not be solely influenced by the perceptions of the severity of the disorder, but also by the psychosocial and practical consequences of treatment[168]. In particular, perceived self-efficacy (parental belief in ability to occlude the child) and

perceived response efficacy (perceived effectiveness of eye occlusion) were positively associated with concordance[221]. However, extrapolation of these findings of studies of children with other forms of amblyopia may be misleading[222].

The management of amblyopia in children with congenital cataract differs from that of other forms of amblyopia in terms of its intensity and, usually, age at commencement. Children with congenital cataract will be operated on at a much earlier age, when occlusion is easier, and some forms of strabismus may not involve surgery at all. Parental experiences, therefore of caring for their child with congenital cataract may possibly involve more prior invasive procedures making them more resilient to the potential stresses of occlusion and more aware of its importance to visual outcome.

A prospective study of children with congenital cataract using an occlusion dose monitor as described earlier and the new Amblyopia Treatment Index[167] and/or the Protective Motivation Theory Questionnaire[221] would enable the delineation of factors likely to improve concordance. A sufficient sample size to enable analyses by age subgroups would be important. Finally, the incorporation of qualitative methods such as interviews would considerably deepen understanding of parental behaviour and attitudes.

Identification of underlying causes of difficulties with occlusion may facilitate a change of emphasis in the management of children with congenital cataract.

Increased orthoptic input, with more active emphasis on visual acuity improvements and consultations with parents who have successfully occluded their child has been recommended for children with other forms of amblyopia[221]and may also be appropriate for children with congenital cataract. Providing written information on the critical period, importance of occlusion and potential negative effects of not treating amblyopia for parents has also shown to be beneficial in improving concordance[159].

6.4 Quality of Life

Two thirds of the PedsQL4.0™ questionnaires were completed at home and although it was emphasised to parents not to influence their child's answers, the extent to which this was adhered to is impossible to gauge. Thus the comparisons of parental and child responses may not involve fully independent observations but the impact of dependency would be to *reduce* child-parent disparity.

Other studies have reported functional visual outcomes in children with ophthalmic disorders using questionnaires[223], or have attempted to address the impact of specific treatment (especially occlusion) on psychosocial aspects of families, but the present study is the first in which health related quality of life has been measured. The PedsQL4.0™ had some limitations as 8(20%) children did not attempt to complete it. This may have been because of poor vision, however at least 3 children had some cognitive disability disorder which together with impaired acuity may have affected their ability to complete the questionnaire. The other children may have failed to complete the questionnaire because the parents were unconcerned about their child completing a questionnaire but were sufficiently interested in participating in the study to complete the questionnaire themselves. This would have occurred in any child self complete HRQOL questionnaire and may not be a problem with the PedsQL4.0™ per se. In other studies using the PedsQL4.0™, the researcher supervised completion[224] and all of the children completed their questionnaires. This concurs with experience in

the present study in which all 14 supervised children completed the questionnaire. Supervised administration of the PedsQL4.0™ e.g. by completion in clinic, is likely to be more effective than postal studies. Of the questionnaires that were completed, there were minimal missing responses suggesting that parents and children are able to provide good quality data regarding the child's health related HRQOL using the PedsQL4.0™. The PedsQL4.0™ internal consistency reliabilities exceeded the group comparison minimal limit of 0.70 and exceeded or approached 0.90 recommended for individual analysis[194]. Furthermore, the measure shows a good range with no floor effects in either the child self or parental reports. However there was a ceiling effect in the physical summary scores for both child and parent scores and it may be that despite possible visual impairment these children are relatively well physically and an 8 item score does not provide an adequate assessment of their HRQOL in this respect. This has found to be the case in other studies of other disorders using the PedsQL4.0™[224]. However, this was found in only one of the scores and on balance, the PedsQL4.0™ is probably still a good tool to use. Overall, experience of the use of the PedsQL4.0™ in the present study supports its further work for the assessment of HRQOL in other groups of children with congenital cataract.

There are no comparable studies of the HRQOL of children with ophthalmic disorders, however, the PedsQL scores of children with congenital cataract in the present study were comparable to those reported by children with notably more debilitating or life threatening diseases such as childhood cancers[224] and

rheumatological disease[225]. Furthermore, the scores of the children with congenital cataract and their parents were consistently lower in all modalities compared to children with acute lymphoblastic leukaemia[224]. This important finding is unexpected and has implications for ophthalmologists regarding how they view the impact of congenital cataract on the HRQOL of their patients. The similarity of the score may be a reflection of the chronicity of all the disorders rather than specifics of the disease per se. This highlights the need for the development of a paediatric, vision related QOL measure which would help determine the impact of specific visual problems on QOL.

Comparison of summary physical health score mean with psychosocial health scores in children with congenital cataract implies that although the children were generally physically well, they experienced compromise to their psychosocial health. This may partly reflect on going concern about visual impairment and vulnerability to late effects. The findings need to be substantiated elsewhere but imply that psychosocial support and careful advice about long term visual prognosis is important long after active treatment ends, even if affected children are generally in good health.

Parental scores showed poor agreement with the child's self reported scores (Bland Altman limits of agreement of the Total Scale Score: parental scores may vary from 36.7 (CI= 25.8 to 47.6) points below and 34 (CI=23.2 to 45.0) points above that of the child). A similar level of disagreement was found between child

and parent in the psychosocial and physical summary scores. This emphasises the importance of assessing HRQOL using measures that both child and parent, or other proxy can complete wherever possible. There may be situations in which the parent's view is all that is available, such as when the child is cognitively impaired and unable to report for herself or other instances such as when a child is too ill or too young to complete questionnaires. Importantly, management decisions about surgery for congenital cataract are mainly in very young children and at an age when a child may be unable to complete a questionnaire.

The differences in agreement are perhaps due to parent's perceptions of real difficulties experienced by the children or their expectation of future problems. Parents may not know about the child's HRQOL in certain situations such as in school and some children may be adept at hiding their feelings from their parents. Parents also have a wider breadth of experiences and may therefore anticipate problems and have different expectations based on other children that are outside those conceivable by the child. Parents own mental health will also be reflected in their reporting on behalf of the children. They may be better at recognising the HRQOL in certain situations such as the impact on the family, sibling relationships and school progress. Children differ from adults in their understanding of health, their beliefs about medicines and hospitals and the causes of illness. However, concordance between the child and parental scores is a necessary requirement for determining the reliability of new measures and a disagreement in the scores raises questions as to whose view is 'correct'[227]. One

solution is to regard each of the assessments as valid and contributing to the whole picture of that child's HRQOL.

Furthermore, agreement between child and parental reports would be expected to be better for the physical summary scores (observable) compared with psychosocial health (less observable) as generally is the case[195] but this was not echoed in this study. This may be because for sighted parents (in the majority of cases) the thought of losing sight may appear to have more impact on physical functioning than actually occurs in children who have always had poor sight. Parents may also take on an overly protective role and do things for their children or prevent their children from doing things that they can probably manage by themselves.

The study size was too small to allow analysis of children in different age subgroups. There are also limitations in comparing HRQOL using only one generic tool and these findings may be reflecting unique features of the PedsQL4.0^{TM}. Studies including groups of healthy children as comparisons would determine PedsQL4.0^{TM}'s validity and studies of the same group would help assess responsiveness of the PedsQL4.0^{TM} in different situations such as pre and post ophthalmic treatment. Further studies in children with eye conditions using different tools would also be informative. However the greatest need is for a robust paediatric vision related quality of life instrument.

These findings should be of value to a parent of a child with newly diagnosed congenital cataract. Despite the need for life long follow up in the majority of cases, possible stressful amblyopia treatment and surgical interventions, children and parents on the whole report a reasonable quality of life. The results also have implications for clinics of ophthalmic patients. The PedsQL 4.0^{TM} could be used as an adjunct to a consultation. Children and their parents who find it difficult to express themselves may find it easier to complete a questionnaire first or after a clinic appointment. Furthermore, the use of a HRQOL tool can provide an invaluable long-term outcome measure of for example, a clinical intervention and be used longitudinally to assess the development of one child. HRQOL instruments could also supplement the application of a child when being statemented for special needs.

The future of HRQOL measures in paediatric ophthalmology lies in the development of a vision related paediatric HRQOL tool. Ophthalmic conditions can carry high morbidity and are associated with other medical conditions. A specific tool would be specific and responsive to children with ophthalmic conditions. It should ideally have parallel child and parental sections and cover a range of ages. It should also be patient centred and ensure that the measure is evaluating quality of life and not general health status[172]. In addition there should be equal emphasis on psychosocial outcomes and the impact of visual impairment as well as functional outcomes, the former being lacking in most currently available adult tools[190]. At present the PedsQL offers a quick, reliable,

easy and valuable measure of HRQOL of children with congenital cataract. Importantly, for children with congenital cataract, when intervention starts at a very young age, it can be used to assess the HRQOL of children as young as 2 years.

7. CONCLUSION

Congenital cataract is a priority of Vision 2020[3], the international programme for the elimination of avoidable blindness, reflecting both the good potential for treatment and the consequences for the child and family if left untreated.

The research reported in this thesis was undertaken to determine the visual acuity and postoperative complications of children with congenital cataract. Furthermore, the predictors of visual acuity and postoperative complications were identified and quantified to inform secondary and tertiary management, as much necessary population-based data in this area have been lacking to date.

61% of children with bilateral cataracts in the present study achieved a visual acuity, by eye, of at least 6/18: commonly considered the level of vision at which children can be educated at mainstream schools with minimum extra help indicating that children with congenital cataract on the whole have good functional outcomes. The worst median visual acuity (6/48) was recorded in the cataractous eyes of children with unilateral cataract who underwent surgery, suggesting that despite intervention in these children, the visual outcome is still poor. Thus appropriate treatment (early surgery and aggressive amblyopia treatment) for many children may not predictably provide the useful "spare" eye that clinicians often propose as the main reason for treating children with unilateral disease. Nevertheless there has, overall been an improvement over time in visual outcomes in children with both unilateral and bilateral congenital

cataract, reflecting improved detection rates, surgical techniques and postoperative amblyopia treatment regimes.

From the present work, timely detection and thus treatment of children with congenital cataract, i.e. within the critical period so as to mitigate against amblyopia, appear to be essential factors in determining good post-operative visual acuity. This supports the continuation of the current infant screening programme in the UK, improved training of screeners (paediatricians and general practitioners undertaking the newborn and 6 week examinations respectively) as well as increased parental and other health professionals' awareness of the importance of timely referral of children suspected to have visual problems. However, the results of this study also show that early surgery may increase the risk of postoperative glaucoma and other complications. This, paradoxically, may be the price of successful screening programmes in which early detection results in earlier surgery. Further work is required to delineate precisely the optimum period of time by which surgery can be delayed to reduce the risk of glaucoma and other postoperative complications without increasing the risk of amblyopia. Prevention of such postoperative complications is essential as currently treatment options are limited and the visual results often poor.

The findings of this study support the continued practice of lensectomy/vitrectomy and contact lens correction as the primary procedure of choice in infants. As IOL implantation is now becoming increasingly more common, and has become the

standard therapy in treatment of children >2 years old with cataract, it is important that further studies, preferably randomised controlled trials, are undertaken to compare the two techniques in younger children.

Occlusion to prevent amblyopia emerges as an important factor to achieve good visual acuity, and is probably the most critical in children with unilateral cataracts. Therefore this study also aimed to explore the psychosocial impact of amblyopia treatment and to assess if these factors affected occlusion concordance.. Most parents in this study found occlusion difficult with a third considering both their child's behaviour and their relationship with their child had worsened as a result of occlusion. Despite this, no association was found between occlusion concordance and the child's or parent's experience of occlusion and most parents never worried that occlusion was harmful. Further studies to identify the underlying causes of difficulties with occlusion and identification of methods to help parents with occlusion may initiate changing emphasis on the management of children with congenital cataract.

This study also aimed to study the feasibility of assessing the HRQOL of children with congenital cataract and to compare the QOL of these children with children with other diseases. This study shows that parents and children were willing and able to complete self reported questionnaires. The generic health-related quality of life scores of children with congenital cataract in the present study were similar to those reported by children with childhood cancers and rheumatological

disorders. This unexpected finding has implications for ophthalmologists regarding how they view the impact of congenital cataract on their patients and their families. Importantly, as the psychosocial scores of these children were lower than their physical scores, psychosocial support and careful advice about long term visual prognosis, are likely to be important long after active treatment ends. More generally, the findings support the development of paediatric, vision related QOL measures which would help determine the specific impact of visual problems on HRQOL.

8. REFERENCES

1. Gilbert C, Rahi JS, Quinn E. In: Johnson GJ, Minassian DC, Weale R, West SK, eds. *The epidemiology of eye disease*. Second ed. Baltimore: Arnold, 2003: 260-86.

2. Gilbert CE, Foster A. Childhood blindness in the context of VISION 2020 - The Right to Sight. *Bull World Health Organ* 2001;**79**:227-32.

3. World Health Organisation. Global initiative for the elimination of avoidable blindness. WHO/PBL/97.61. 1977. Geneva.

4. Straatsma BR. Clinical trials in the past 25 years and clinical research in the next 25 years. *Am J Ophthalmol* 1999;**127**:72-4.

5. Snell RS, Lemp MA. In: *Clinical anatomy of the eye*. Second ed. Blackwell Science, 1998: 1-20.

6. Paterson CA, Delamere NA. In: Hart WM, ed. *Adler's physiology of the eye*. St Louis, Missouri: Mosby-Year Book, 1992: 348-90.

7. Forrester J, Dick A, McMenamin P, Lee W. In: *The Eye:Basic sciences in practice*. Londom: WB Saunders Company Ltd, 1996: 137-98.

8. Taylor D. Congenital cataract: the history, the nature and the practice. The Doyne Lecture. *Eye* 1998;**12**:9-36.

9. Francis PJ, Berry V, Moore AT, Bhattacharya S. Lens biology: development and human cataractogenesis. *Trends in Genetics* 1999;**15**:191-6.

10. Francis PJ, Berry V, Bhattacharya S, Moore AT. Genetics of childhood cataract. *J Med Genetics* 2000;**37**:481-8.

11. Moore AT. Understanding the molecular genetics of congenital cataract may have wider implications for age related cataract. *BMJ* 2004;**88**:2-3.

12. Lambert SR, Drack AV. Infantile cataracts. *Surv Ophthalmol* 1996;**40**:427-58.

13. Moore BD. Pediatric cataracts-diagnosis and treatment. *Optometry and Visual Science* 1994;**71**:168-73.

14. Parks MM, Johnson DA, Reed GW. Long-term Visual Results and Complications in Children with Aphakia. *Ophthalmology* 1993;**100**:826-41.

15. Horton JC. In: Hart WM, ed. *Adler's physiology of the eye*. 9th ed. St Louis: Mosby Year Book, 1992: 728-72.

16. Wiesel TN, Hubel TH. Single cell responses in striate cortex of kittens deprived of vision in one eye. *J Neurophysiol* 1963;**26**:1003-17.

17. Von Noorden GK. Amblyopia: A multidisciplinary approach. *Invest Ophthalmol Vis Sci* 1985;**26**:1704-16.

18. Hubel TH, Wiesel TN. Binocular interaction in striate cortex of kittens reared with artificial squint. *J Neurophysiol* 1965;**28**:1041-59.

19. Von Noorden GK, Khodadoust A. Retinal haemorrhage in newborns and organic amblyopia. *Arch Ophthalmol* 1973;**89**:91-3.

20. Taylor D, Rice NSC. Congenital cataract, a cause of preventable child blindness. *Arch Dis Child* 1982;**57**:165-7.

21. Vaegan, Taylor D. Critical period for deprivation amblyopia in children. *Trans Ophthalmol Soc U.K.* 1979;**99**:432-9.

22. Mitchell DE, Cynader M, Movshon JA. Recovery from the effects of monocular deprivation in kittens. *J Comp Neurol* 1977;**176**:53-64.

23. Fulton AB, Dobson V, Salem D, Mar C, Petersen RA, Hansen RM. Cycloplegic refractions in infants and young children. *Am J Ophthalmol* 1980;**90**:239-47.

24. Wiesel TN. Postnatal development of the visual cortex and the influence of environment. *Nature* 1982;**299**:583-91.

25. Rahi JS, Cable N, British Childhood Visual Impairment Study Group. Severe visual impairment and blindness in children in the UK. *Lancet* 2003;**362**:1359-65.

26. Foster A, Gilbert C. In: Taylor D, ed. *Paediatric Ophthalmology*. 2nd ed. London: Blackwell Science, 1997: 3-12.

27. Smith AF, Smith JG. The economic burden of global blindness: a price too high! *Br J Ophthalmol* 1996;**80**:276-7.

28. Rahi JS, Gilbert CE. In: Taylor D, Hoyt CS, eds. *Paediatric Ophthalmology*. Third ed. London: Elsevier, 2004.

29. Foster A, Gilbert C, Rahi J. Epidemiology of cataract in childhood: A global perspective. *J Cataract Refract Surg* 1997;**23**:601-4.

30. Foster A, Gilbert C. Epidemiology of childhood blindness. *Eye* 1992;**6**:173-6.

31. Abrahamsson M, Magnusson G, Sjostrom A, Popvic Z, Sjostrand J. The occurrence of congenital cataract in western Sweden. *Acta Ophthalmol Scand* 1999;**77**:578-80.

32. Wirth MG, Russell-Eggitt IM, Craig JE, Elder JE, Mackey DA. Aetiology of congenital and paediatric cataract in an Australian population. *Br J Ophthalmol* 2002;**86**:782-6.

33. James LM. Maps of birth defects occurrence in the U.S., Birth Defects Monitoring Program (BDMP)/CPHA, 1970-1987. *Teratology* 1993;**48**:551-646.

34. Stoll C, Alembik Y, Dott B, Roth MP. Epidemiology of congenital eye malformations in 131,760 consecutive births. *Ophthalmic Paediatrics and Genetics* 1992;**13**:179-86.

35. Jensen H, Goldschmidt E. Visual acuity in Danish school children. *Acta Ophthalmol* 1986;**64**:187-91.

36. Stayte M, Reeves B, Wortham C. Ocular and vision defects in preschool children. *Br J Ophthalmol* 1993;**77**:228-32.

37. Kohler L, Stigmar G. Vision screening of four-year-old children. *Acta Paediatr.Scand.* 1973;**62**:17-27.

38. Stewart-Brown S, Haslum MN. Partial sight and blindness in children of the 1970 birth cohort at 10 years of age. *J Epidemiol Community Health* 1988;**42**:17-23.

39. Hu D. Prevalence and mode of inheritance of major genetic eye diseases in China. *Journal of Medical Genetics* 1987;**24**:584-8.

40. Chirambo MC, Tielsch JM, West KP, Jr., Katz J, Tizazu T, Schwab L et al. Blindness and visual impairment in southern Malawi. *Bull World Health Organ* 1986;**64**:567-72.

41. Brilliant LB, Pokhrel RP, Grasset NC, Lepkowski JM, Kolstad A, Hawks W et al. Epidemiology of blindness in Nepal. *Bull World Health Organ* 1985;**63**:375-86.

42. Rahi JS, Williams C, Bedford H, Elliman D. Screening and surveillance for ophthalmic disorders and visual deficits in children in the United Kingdom. *Br J Ophthalmol* 2001;**85**:257-9.

43. Auld R. The management of paediatric cataracts: a retrospective study. *Br Orthopt J* 1989;**46**:30-9.

44. Wright K.W., Christensen L.E, Noguchi B.A. Results of late surgery for presumed congenital cataracts. *Am J Ophthalmol* 1992;**114**:409-15.

45. Committee on Practice and Ambulatory Medicine, American Association of Certified Orthoptists, American Association for Pediatric Ophthalmology and Strabismus, American Academy of Ophthalmology. Eye examination in infants, children, and young adults by pediatricians. *Pediatrics* 2003;**111**:902-7.

46. Rahi JS, Lynn R. A survey of paediatricians' practice and training in routine infant eye examination. *Arch Dis Child* 1998;**78**:364-6.

47. Rahi JS, Dezateux C, for the British Congenital Cataract Interest Group. National cross-sectional study of detection of congenital and infantile cataract in the United Kingdom: role of screening and surveillance. *BMJ* 1999;**318**:362-5.

48. Eckstein M, Vijayalakshmi P, Gilbert C, Foster A. Randomised clinical trial of lensectomy versus lens aspiration and primary capsulotomy for children with bilateral cataract in south India. *Br J Ophthalmol* 1999;**83**:524-9.

49. Vasavada AR, Trivedi RH. Role of optic capture in congenital cataract and intra-ocular lens surgery in children. *J Cataract Refract Surg* 2000;**26**:824-31.

50. Basti S, Aasuri MK, Reddy MK, Preetam P, Reddy S, Gupta S et al. Heparin surface modified intra-ocular lenses in pediatric cataract: prospective randomised study. *J Cataract Refract Surg* 1999;**25**:782-7.

51. Myers VS, Gridlewski N, Quinn GE, Dobson V. Distance and near visual acuity, contrast sensitivity and visual fields of 10 year old children. *Arch Ophthalmol* 1999;**117**:94-9.

52. Morad Y, Werker E, Nemet P. Visual acuity tests using chart, line and singlr optotype in healthy and amblyopic children. *J AAPOS* 1999;**3**:94-7.

53. Manny RE, Hussein M, Gwiazda J, Marsh-Tootle W, COMET Study Group. Repeatibility of ETDRS visual acuity in children. *Invest Ophthalmol Vis Sci* 2003;**44**:3294-300.

54. Simmers AJ, Gray LS, Spowart K. Screening for amblyopia: a comparison of paediatric letter tests. *Br J Ophthalmol* 1997;**81**:465-9.

55. Holmes JM, Beck RW, Repka MX, Leske DA, Kraker RT, Clifford Blair R et al. The amblyopia treatment study visual acuity testing protocol. *Arch Ophthalmol* 2001;**119**:1345-53.

56. Kheterpal S, Jones H S, Auld R, Moseley M J. Reliability of visual acuity in children with reduced vision. *Ophthal Physiol Opt* 1996;**16**:447-9.

57. Harrad R. Modulation of amblyopia therapy following early surgery for unilateral congenital cataracts. *Br J Ophthalmol* 1995;**79**:793.

58. Maurer D, Lewis TL. In: Simons K, ed. *Early visual development, normal and abnormal.* 1st ed. New York: Oxford University Press, 1993: 454-84.

59. Hing SJ, Speedwell L, Taylor D. Lens surgery in infancy and childhood. *Br J Ophthalmol* 1990;**74**:73-7.

60. Harwerth RS, Smith III EL, Paul AD, Crawford MLJ, Von Noorden GK. Functional effects of visual form deprivation in monkeys. *Invest Ophthalmol Vis Sci* 1991;**32**:2311-27.

61. Elston JS, Timms C. Clinical evidence for the onset of the sensitive period in infancy. *Br J Ophthalmol* 1992;**76**:327-8.

62. Birch EE, Stager DR. The critical period for surgical treatment of dense congenital unilateral cataract. *Invest Ophthalmol Vis Sci* 1996;**37**:1532-8.

63. Birch EE, Swanson WH, Stager DR, Woody M, Everett M. Outcome after very early treatment of dense congenital unilateral cataract. *Invest Ophthalmol Vis Sci* 1993;**34**:3687-99.

64. Birch EE, Stager DR, Leffler J, Weakley D. Early treatment of congenital unilateral cataract minimizes unequal competition. *Invest Ophthalmol Vis Sci* 1998;**39**:1560-6.

65. Lloyd IC, Dowler JGF, Kriss A, Speedwell L, Thompson DA, Russell-Eggitt I et al. Modulation of amblyopic therapy following early surgery for unilateral congenital cataracts. *Br J Ophthalmol* 1995;**79**:802-6.

66. Gregg FM, Parks MM. Stereopsis After Congenital Monocular Cataract Extraction. *Am J Ophthalmol* 1992;**114**:314-7.

67. Drummond G.T, Scott WE, Keech RV. Management of monocular congenital cataracts. *Arch Ophthalmol* 1989;**107**:45-51.

68. Brown AM, Yamamoto M. Visual acuity in newborn and preterm infants measured with grating acuity cards. *Am J Ophthalmol* 1985;**102**:245-53.

69. Taylor DSI, Wright LA, Amaya L, Cassidy L, Nischal K, Russell-Eggitt IM. Should we aggressively treat unilateral congenital cataracts? *Br J Ophthalmol* 2001;**85**:1120-6.

70. Scheie HG. Aspiration of congenital or soft cataracts: a new technique. *Am J Ophthalmol* 1960;**50**:1048-56.

71. Wheeler DT, Stager DR, Weakley D. Endophthalmitis following pediatric intraocular surgery for congenital cataracts and congenital glaucoma. *J Pediatr Ophthalmol Strabismus* 1992;**29**:139-41.

72. Ahmadieh H, Javadi MA, Ahmady M, Karimian F, Einollahi B, Zare M et al. Primary capsulotomy, anterior vitrectomy, lensectomy, and posterior chamber lens implantation in children: limbal versus pars plana. *J Cataract Refract Surg* 1999;**25**:768-75.

73. Wilson ME. Intraocular lens implantation: has it become the standard of care for children? *Ophthalmology* 1996;**103**:1719-20.

74. Young TL, Bloom JN, Ruttum M, Sprunger DT, Weinstein JM on behalf of the AAPOS research committee. The IOLAB,Inc pediatric intra-ocular lens study. *J AAPOS* 1999;**3**:295-302.

75. Zwaan J, Mullaney PB, Awad A, Al-Mesfer S, Wheeler DT. Pediatric intaocular lens implantation. *Ophthalmology* 1998;**105**:112-9.

76. Gimbel HV, Basti S, Ferensowicz M, DeBroff BM. Results of Bilateral Cataract Extraction with Posterior Chamber Intraocular Lens Implantation in Children. *Ophthalmology* 1997;**104**:1737-43.

77. Kora Y, Inatomi M, Fukado Y, Marumori M, Yaguchi S. Long-term study of children with implanted intra-ocualr lenses. *J Cataract Refract Surg* 1992;**18**:485-8.

78. Malukiewicz-Wisniewska G, Kaluzny J, Lesiewska-Junk H, Elkis I. Intra-ocular lens implantation in children and youth. *J Pediatr Ophthalmol Strabismus* 1999;**36**:129-33.

79. Gimbel HV, Ferensowicz M, Raanan M, DeLuca M. Implantation in children. *J Pediatr Ophthalmol Strabismus* 1993;**30**:69-79.

80. Crouch ER, Pressman SH. Posterior chamber intra-ocular lenses: long-term results in pediatric cataract patients. *J Pediatr Ophthalmol Strabismus* 1995;**32**:210-8.

81. Wheeler DT, Mullaney PB, Awad A, Al-Mesfer S, Al-Nahdi T, Zwaan J. Pediatric IOL implantation: the KKESH experience. *J Pediatr Ophthalmol Strabismus* 1997;**34**:341-6.

82. Simons BD, Siatkowski RM, Schiffman JC, Flynn J, Capo H, Munoz M. Surgical technique, visual outcome, and complications of pediatric intra-ocular lens implantation. *J Pediatr Ophthalmol Strabismus* 1999;**36**:118-24.

83. Ainsworth JR, Cohen S, Levin AV, Rootman DS. Pediatric cataract management with variations in surgical technique and aphakic optical correction. *Ophthalmology* 1997;**104**:1096-101.

84. Rice NSC. Closed microsurgery in the treatment of congenital cataracts. *Dev Ophthalmol* 1985;**11**:153-5.

85. Chrousos G.A., Parks MM, O'Neill J.F. Incidence of chronic glaucoma, retinal detachment and secondary membrane surgery in pediatric aphakic patients. *Ophthalmology* 1984;**91**:1238-41.

86. Keech RV, Tongue AC, Scott WE. Complications after surgery for congenital and infantile cataracts. *Am J Ophthalmol* 1989;**108**:136-41.

87. Chandler PA. Surgery of the lens in infancy and childhood. *Arch Ophthalmol* 1951;**45**:125-38.

88. Asrani SG, Wilensky JT. Glaucoma after congenital catarct surgery. *Ophthalmology* 1995;**102**:863-7.

89. Mills M, Robb R. Glaucoma following childhood cataract surgery. *J Pediatr Ophthalmol Strabismus* 1994;**31**:355-60.

90. Magnusson G, Abrahamsson M, Sjostrand J. Glaucoma following congenital cataract surgery:An 18-year longitudinal follow-up. *Acta Ophthalmol Scand* 2000;**78**:65-70.

91. Egbert JE, Wright MM, Dahlhauser KF, Keithahn MAZ, Letson RD, Sumers C. A prospective study of ocular hypertension and glaucoma after pediatric cataract surgery. *Ophthalmology* 1995;**102**:1098-101.

92. Johnson CP, Keech RV. Prevalence of glaucoma after surgery for PHPV and infantile cataracts. *J Pediatr Ophthalmol Strabismu* 1996;**33**:14-7.

93. Vishwanath M, Cheong-Leen R, Taylor D, Russell-Eggitt IM, Rahi JS. Is early surgery for congenital cataract a risk factor for glaucoma? *Br J Ophthalmol* 2004;**88**:905-10.

94. Russell-Eggitt IM, Zamiri P. Review of aphakic glaucoma after surgery for congenital cataract. *J Cataract Refract Surg* 1997;**23**:664-8.

95. Wilson ME, Bluestein E, Wang X-H. Current trends in the use of intra-ocular lenses in children. *J Cataract Refract Surg* 1994;**20**:579-83.

96. Wilson ME, Elliot L, Johnson B, Peterseim M, Rah S, Werner L et al. Acrysof Acrylic Intraocular Lens Implantation in Children:Clinical Indications of Biocompatibility. *J AAPOS* 2003;**5**:377-80.

97. Lundvall A. ZC. Complications after early surgery for congenital cataracts. *Acta Ophthalmol Scand* 1999;**77**:677-80.

98. Rabiah PK. Frequency and predictors of glaucoma after pediatric cataract surgery. *Am J Ophthalmol* 2004;**137**:30-7.

99. Asrani SG, Freedman SF, Hasselblad V, Buckley EG, Egbert JE, Dahan E et al. Does primary intraocular lens implantation prevent aphakic glaucoma in children. *J AAPOS* 1999;**3**:33-9.

100. Lambert SR, Lynn M, Drews-Botsch C, Loupe D, Plager DA, Medow NB et al. A comparison of grating visual acuity, strabismus and reoperation outcomes among children with aphakia and pseudophakia after unilateral cataract surgery during the first six months of life. *J AAPOS* 2001;**5**:70-5.

101. McClatchey SK, Dahan E, Maselli E, Gimbel HV, Wilson ME, Lambert SR et al. A comparison of the rate of refractive growth in pediatric aphakic and pseudophakic eyes. *Ophthalmology* 2002;**107**:118-22.

102. O'Keefe M, Fenton S, Lanigan B. Visual outcomes and complications of posterior chamber intraocular lens implantation in the first year of life. *J Cataract Refract Surg* 2002;**27**:2006-11.

103. Plager DA, Yang S, Neely D, Sprunger DT, Sondhi N. Complications in the first year following cataract surgery with and without IOL in infants and older children. *J AAPOS* 2002;**6**:9-14.

104. Papadopoulos M, Khaw PT. Meeting the challenge of glaucoma after paediatric cataract surgery. *Eye* 2003;**17**:1-2.

105. Kirwan JF, Shah P, Khaw PT. Diode laser cyclophotocoagulation: role in the management of refractory pediatric glaucomas. *Ophthalmology* 2002;**109**:316-23.

106. Mandal AK, Bagga H, Nuthethi R, Gothwal VK, Nanda AK. Trabeculectomy with and without Mitomycin-C for pediatric glaucoma in aphakia and pseudophakia following congenital cataract surgery. *Eye* 2003;**17**:53-62.

107. Azuara-Blanco A, Wilson RP, Spaeth GL, Schmidt CM, Augsburger JJ. Filtration procedures supplemented with mitomycin C in the management of childhood glaucoma. *Br J Ophthalmol* 1999;**83**:151-6.

108. McCleod D. Congenital cataract surgery: a retinal surgeon's viewpoint. *Aust J Ophthalmol.* 1986;**14**:79-84.

109. Toyofuku H, Hirose T, Schepens CL. Retinal detachment following congenital cataract surgery. *Arch Ophthalmol* 1980;**98**:669-75.

110. Good WV, Hing S, Irvine AR, Hoyt C.S., Taylor D. Postoperative endophthalmitis in children following cataract surgery. *J Pediatr Ophthalmol Strabismus* 1990;**27**:283-5.

111. Hosal B.M., Biglan AW. Risk factors for secondary membrane formation after removal of pediatric cataract. *J Cataract Refract Surg* 2002;**28**:302-9.

112. Jensen AA, Basti S, Greenwald MJ, Mets MB. When may the posterior capsule be preserved in pediatric intraocular lens surgery. *Ophthalmology* 2002;**109**:324-8.

113. BenEzra D, Cohen E. Posterior Capsultectomy in Pediatric Cataract Surgery. *Ophthalmology* 1997;**104**:2168-74.

114. Plager DA, Lipsky SN, Snyder SK, Sprunger DT, Ellis FD, Sondhi N. Capsular management and refractive error in pediatric intraocular lenses. *Ophthalmology* 1997;**104**:600-7.

115. Metge P, Cohen H, Chemila JF. Intercapsular implantation in children. *Eur J Implant Ref Surg* 1990;**2**:319-23.

116. Alexandrakis G, Peterseim M, Wilson ME. Clinical outcomes of pars plana capsulotomy with anterior vitrectomy in pediatric cataract surgery. *J AAPOS* 2002;**6**:163-7.

117. Ram J, Brar GS, KAushil S, Gupta A, Gupta A. Role of posterior capsulotomy with vitrectomy and intraocular lens design and material in reducing posterior capsular opacification after pediatric cataract surgery. *J Cataract Refract Surg* 2003;**29**:1579-84.

118. Tassignon MJBR, De Groot V, Vrensen GFJM. Bag-in-the-lens implantation of intraocular lenses. *J Cataract Refract Surg* 2002;**28**:1182-8.

119. Awner S, Buckley EG, DeVaro JM, Seaber JH. Unilateral pseudophakia in children under 4 years. *J Pediatr Ophthalmol Strabismus* 1996;**33**:230-6.

120. Brady KM, Atkinson CS, Kilty LA, Hiles DA. Cataract surgery and intraocular lens implantation in children. *Am J Ophthalmol* 1995;**120**:1-9.

121. Sinskey RM, Stoppel JO, Amin P. Long-term results of intra-ocular lens implantation in pediatric patients. *J Cataract Refract Surg* 1993;**19**:405-8.

122. Lloyd IC. Intra-ocular lens implantation in infants. *Clinical and Experimental Ophthalmology* 2000;**28**:338-40.

123. Rogers v Whittaker. (67 ALJR 47). 1992. Australia High Court.

124. Christiansen SP, Munoz M, Capo H. Retinal haemorrhage following lensectomy and anterior vitrectomy in children. *J Pediatr Ophthalmol Strabismus* 1993;**30**:24-7.

125. Lenart TD, Drack AV, Tarnuzzer RW, Fernandes A, Lambert SR. Heterochromia after pediatric cataract surgery. *J AAPOS* 2000;**4**:40-5.

126. Rao SK, Ravishankar K, Sitalakshmi G, Ng JSK, Yu C, Lam DSC. Cystoid macular edema after pediatric intraocular lens implantation:fluorescein

angioscopy results and literature review. *J Cataract Refract Surg* 2001;**27**:432-6.

127. Parks MM. Management of the posterior capsule in congenital cataracts. *J Pediatr Ophthalmol Strabismus* 1984;**21**:114-7.

128. Amaya L, Speedwell L, Taylor D. Contact lens for infant aphakia. *Br J Ophthalmol* 1990;**74**:150-4.

129. Ma JJK, Morad Y, Mau E, Brent HP, Barclay R, Levin AV. Contact lenses for the treatment of pediatric cataracts. *Ophthalmology* 2003;**110**:299-305.

130. Moore BD. Pediatric Aphakic Contact Lens Wear: Rates of Successful Wear. *J Pediatr Ophthalmol Strabismus* 1993;**30**:253-8.

131. de Brabander J, Kok JHC, Nuijts RMMA, Wenniger-Prick LJJM. A practical approach to long-term results of fitting silicone contact lenses in aphakic children after congenital cataract. *CLAO J* 2002;**28**:31-5.

132. Peterseim MW, Wilson ME. Bilateral intraocular lens implantation in the pediatric population. *Ophthalmology* 2000;**107**:1261-6.

133. Cassidy L, Rahi JS, Nischal K, Russell-Eggitt IM, Taylor DSI. Outcome of lens aspiration and intra-ocular lens implantation in children aged 5 years and under. *Br J Ophthalmol* 2001;**85**:540-2.

134. Wilson ME, Bartholomew LR, Trivedi RH. Pediatric cataract surgery and intraocular lens implantation. Practice styles and preferences of the 2001

ASCRS and AAPOS memberships. *J Cataract Refract Surg* 2003;**29**:1811-20.

135. Burke JP, Willshaw HE, Young JDH. Intra-ocular lens implants for uniocular cataracts in childhood. *Br J Ophthalmol* 1989;**73**:860-**4**.

136. Knight-Nanan D, O'Keefe M, Bowell R. Outcome and complications of intra-ocular lenses in children with cataract. *J Cataract Refract Surg* 1996;**22**:730-6.

137. Lambert SR, Buckley EG, Plager DA, Medow NB, Wilson ME. Unilateral intra-ocular lens implantation during the first six months of life. *J AAPOS* 1999;**3**:344-9.

138. Lesueur LC, Arne JL, Chapotot EC, Thouvenin D, Malecaze F. Visual outcome after paediatric cataract surgery: is age a major factor? *Br J Ophthalmol* 1998;**82**:1022-5.

139. Markham RHC, Bloom PA, Chandna A, Newcomb EH. Results of intra-ocaulr lesn implantation in paediatric aphakia. *Eye* 1992;**6**:493-8.

140. Sinskey RM, Amin P, Lingua R. Cataract extraction and intraocular lens implantation in an infant with a monocular cataract. *J Cataract Refract Surg* 1994;**20**:651.

141. Vasavada AR, Chauhan H. Intraocular lens implantation in infants with congenital cataract. *J Cataract Refract Surg* 1994;**20**:592-8.

142. Mullner-Eidenbock A, Amon M, Moser E, Kruger A, Abela C, Schlemmer Y et al. Morphological and functional results of Acrysof intraocular lens implantation in children. *J Cataract Refract Surg* 2003;**29**:285-93.

143. Trivedi RH, Wilson ME. Single piece acrylic intraocular lens implantation in children. *J Cataract Refract Surg* 2003;**29**:1738-43.

144. Wilson ME, Apple DJ, Bluestein E, Wang X-H. Intraocular lenses for pediatric implantation:biomaterials, designs and sizing. *J Cataract Refract Surg* 1994;**20**:584-91.

145. McClatchey SK, Parks MM. Myopic Shift after cataract removal in childhood. *J Pediatr Ophthalmol Strabismus* 1997;**34**:88-95.

146. Filtcroft DI, Knight-Nanan D, Bowell R, Lanigan B, O'Keefe M. Intraocular lenses in children: changes in axial length, corneal curvature, and refraction. *Br J Ophthalmol* 1998;**83**:265-9.

147. Kashani AA. Refractive Changes after Lens Implantation in Childhood. *Ophthalmology* 1998;**105**:1571.

148. McClatchey SK. Intraocular lens calculator for childhood cataract. *Journal of Cataract Refraction Surgery* 1998;**24**:1125-9.

149. Lorenz B, Worle J, Friedl N, Hasenfratz G. Ocular growth in infant aphakia. *Ophthalmic Paediatrics and Genetics* 1993;**14**:177-88.

150. Wilson ME, Englert JA, Greenwald MJ. In-the-bag secondary intraocular lens implantation in children. *J AAPOS* 1999;**3**:350-5.

151. Morgan KS, Stephenson GS, McDonald MB, Kaufman HE. Epikeratophakia in children. *Ophthalmology* 1984;**91**:780-4.

152. Simons K, Preslan M. Natural history of amblyopia untreated owing to lack of compliance. *Br J Ophthalmol* 1999;**83**:582-7.

153. Clarke MP, Wright CM, Hrisos S, Anderson JD, Henderson J, Richardson SR. Randomised controlled trial of treatment of unilateral visual impairment detected at preschool vision screening. *BMJ* 2003;**327**:1251-60.

154. Bradford G, Keech RV, Scott W. Factors affecting visual outcome after surgery for bilateral congenital cataracts. *Am J Ophthalmol* 1994;**117**:58-64.

155. Taylor D, Vaegan, Morris R, Rodgers J.E., Warland J. Amblyopia in bilateral infantile and juvenile cataract. Relationship to timing of treatment. *Trans Ophthalmol Soc U.K.* 1979;**99**:170-5.

156. Elsas FJ. Visual Acuity in Monocular Pediatric Aphakia: Does Epikeratophakia Facilitate Occulsion Therapy in Children Intolerant of Contact Lens or Spectacle Wear? *J Pediatr Ophthalmol Strabismus* 1990;**27**:304-9.

157. Smith LK, Thompson JR, Woodruff G, Hiscox F. Factors effecting treatment compliance in amblyopia. *J Pediatr Ophthalmol Strabismus* 2004;**32**:98-101.

158. Newsham D. Parental non-concordance with occlusion therapy. *Br J Ophthalmol* 2000;**84**:957-62.

159. Newsham D. A randomised controlled trial of written information: the effect on parental non-concordance with occlusion therapy. *Br J Ophthalmol* 2002;**86**:787-91.

160. Jeffrey BG, Birch EE, Stager DR, Weakley D. Early binocular visual experience may improve binocular sensory outcomes in children after surgery for congenital unilateral cataract. *J AAPOS* 2001;*5*:209-16.

161. Holmes JM, Kraker RT, Beck RW, Birch EE, Cotter SA, Everett DRW et al. A randomized trial of prescribed patching for treatment of severe amblyopia in children. *Ophthalmology* 2003;**110**:2075-87.

162. Keech RV, Kutschke PJ. Upper age limit for the development of amblyopia. *J Pediatr Ophthalmol Strabismus* 1995;**32**:89-93.

163. Lewis TL, Maurer D, Tytla M, Bowering ER, Brent HP. Vision in the good eye of children treated for unilateral congenital cataract. *Ophthalmology* 1992;**99**:1013-7.

164. Thompson DA, Moller H, Russell-Eggitt I, Kriss A. Visual acuity in unilateral cataract. *Br J Ophthalmol* 1996;**80**:794-8.

165. Smith KH, Baker DB, Keech RV, Adams LW, Rosa RH, Austin CJ et al. Monocular congenital cataracts:psychological effects of treatment. *J Pediatr Ophthalmol Strabismus* 1991;**28**:245-9.

166. Rogers GL, Tishler CL, Tsou BH, Hertle RW, Fellows RR. Visual acuities in infants with congenital cataracts operated on prior to 6 months of age. *Arch Ophthalmol* 1981;**99**:999-1003.

167. Pediatric Disease Investigator Group. Impact of patching and atropine treatment on the child and family in the amblyopia treatment study. *Arch Ophthalmol* 2003;**121**:1625-32.

168. Searle A, Vedhara K, Norman P, Frost A, Harrad R. Compliance with eye patching in children and its psychosocial effects: a qualitative application of protection motivation theory. *Psychology, health & medicine* 2000;**1**:43-54.

169. Foley-Nolan A, McCann A, O'Keefe M. Atropine penalisation versus occlusion as the primary treatment for amblyopia. *Br J Ophthalmol* 1997;**81**:54-57.

170. Aristotle, Translated by Terence Unwin. *Nicomachean Ethics*. Indianapolis Hackett Publishing Company, 1985.

171. Brown P. Quality of life 'getting better'. The Guardian Newspaper Thursday March 14th. 2002.

172. Carr AJ, Higginson IJ. Are quality of life measures patient centred? *BMJ* 2001;**322**:1357-60.

173. Eiser C. Children's quality of life measures. *Arch Dis Child* 1997;**77**:350-4.

174. Jenney ME, Campbell S. Measuring quality of life. *Arch Dis Child* 1997;**77**:347-54.

174a. World Health Organisation. *Constitution of the WHO.* Geneva: WHO, 1958;Annex I

175. Higginson IJ, Carr AJ. Using quality of life measures in the clinical setting. *BMJ* 2001;**322**:1297-300.

176. Carr AJ, Thompson PW, Young A. Do health status measures have a role in rheumatology? *Arthritis and Rheumatism* 1996;**39**:261S.

177. Eiser C, Morse R. A review of measures of quality of life for children with chronic illness. *Arch Dis Child* 2001;**84**:205-11.

178. Wiebe S, Guyatt G, Weaver B, Matijevic S, Sidwell C. Comparative responsiveness of generic and specific quality of life instruments. *Journal of Clinical Epidemiology* 2003;**56**:52-60.

179. Carr AJ, Gibson B, Robinson PG. Is quality of life determined by expectations or experience? *BMJ* 2001;**322**:1240-3.

180. Pal DK. Quality of Life Assessment in Children: A Review of Conceptual and Methodological Issues in Multidimensional Health Status Measures. *Epidemiol Community Health* 1996;**50**:391-6.

181. Crisp J, Ungerer JA, Goodnow JJ. The impact of experience on children's understanding of illness. *J Paediatr Psychol* 1996;**21**:57-72.

182. Bibace R, Walsh ME. Development of children's concepts of illness. *Pediatrics* 1980;**66**:912-7.

183. Addington-Hall, Kalra L. Who should measure quality of life? *BMJ* 2001;**322**:1417-20.

184. Nichols JJ, Mitchell L, Saracino M, Zadnik K. Reliability and validity of refractive error-specific quality of life instruments. *Arch Ophthalmol* 2003;**121**:1289-96.

185. Massof R, Fletcher DC. Evaluation of the NEI visual functioning questionnaire as an interval measure of visual ability in low vision. *Vision Research* 2001;**41**:397-413.

186. Massof R, Rubin GS. Visual Function Assessment Questionnaires. *Surv Ophthalmol* 2001;**45**:531-48.

187. Chan CWN, Wong JCC, Chan KSK, Wong WK, Tam KCK, Pearl SY. Evaluation of quality of life in patients with cataract in Hong Kong. *J Cataract Refract Surg* 2003;**29**:1753-60.

188. Mangione CM LP, Gutierrez PR, Spritzer K, Berry S, Hays RD. Development of the 25-item national eye institute visual function questionnaire. *Arch Ophthalmol* 2001;**119**:1050-8.

189. Javitt JC, Jacobson G, Schiffman RM. Validity and reliability of the cataract type spec:an instrument for measuring outcomes of cataract extraction. *Am J Ophthalmol* 2003;**136**:285-90.

190. Margolis MK, Coyne K, Kennedy-Martin T, Baker T, Schein O, Revicki DA. Vision-specific instruments for the assessment of health related quality of life and visual functioning. *Pharmacoeconomics* 2002;**20**:791-812.

191. The Submacular Surgery Trials Research Group. Responsiveness of the National Eye Institute visual function questionnaire to changes in visual acuity. *Arch Ophthalmol* 2003;**121**:531-9.

192. Mendes F, Schaumberg DA, Navon S, Steinert R, Sugar J, Holland EJ et al. Assessment of visual function after corneal transplantation:the quality of life and psychometric assessment after corneal transplantation (Q-PACT) study. *Am J Ophthalmol* 2003;**135**:785-93.

193. Deramo VA, Cox TA, Syed AB, Lee PP, Fekrat S. Vision related quality of life in people with central retinal vein occlusion using the 25 item national eye institute visual function questionnaire. *Arch Ophthalmol* 2003;**121**:1297-302.

194. Varni JW, Seid M, Kurtin P. PedsQL 4.0:Reliability and validity of the pediatric quality of life inventory version 4.0 generic core scales in healthy and patient populations. *Medical Care* 2001;**39**:800-12.

195. Varni JW, Seid M, Rode CA. The PedsQL:Measurement Model for the Pediatric Quality of Life Inventory. *Medical Care* 1999;**37**:126-39.

196. Rahi JS, Dezateux C, for the British Congenital Cataract Interest Group. Capture-recapture analysis of ascertainment by active surveillance in the British Congenital Cataract Study. *Invest Ophthalmol Vis Sci* 1999;**40**:236-9.

197. Rahi JS, Dezateux C, for the British Congenital Cataract Interest Group. Congenital and infantile cataract in the United Kingdom: underlying or associated factors. *Invest Ophthalmol Vis Sci* 2000;**41**:2108-14.

198. Rahi JS, Botting BJ, and the British Congenital Cataract Interest Group. Ascertainment of children with congenital cataract through the National Congenital Anomaly System in England and Wales. *Br J Ophthalmol* 2001;**85**:1049-51.

199. Rahi JS, Dezateux C, for the British Congenital Cataract Interest Group. Measuring and interpreting the incidence of congenital ocular anomalies: lessons from a national study of congenital cataract in the UK. *Invest Ophthalmol Vis Sci* 2001;**42**:1444-8.

200. Newcombe RG, Duff GR. Eyes or patients? Traps for the unwary in the statistical analysis of ophthalmological studies. *Br J Ophthalmol* 1987;**71**:645-6.

201. Murdoch IE, Morris SS, Cousens SN. People and eyes: statistical approaches in ophthalmology. *Br J Ophthalmol* 1998;**82**:971-3.

202. Bland JM, Altman DG. Cronbach's alpha. *BMJ* 1997;572.

203. Bland JM, Altman DG. Statistical methods for assessing agreement between two methods of clinical measurement. *Lancet* 1986;**1**:307-10.

204. Pediatric Disease Investigator Group. The clinical profile of moderate amblyopia in children younger than 7 years. *Arch Ophthalmol* 2002;120:281-7.

205. Lundvall A, Kugelberg U. Outcome after treatment of congenital bilateral cataract. *Acta Ophthalmol Scand* 2002;**80**:593-7.

206. Magnusson G, Abrahamsson M, Sjostrand J. Changes in visual acuity from 4 to 12 years of age in children operated for bilateral congenital cataracts. *Br J Ophthalmol* 2002;**86**:1385-9.

207. Lundvall A, Kugelberg U. Outcome after treatment of congenital unilateral cataract. *Acta Ophthalmol Scand* 2002;**80**:588-92.

208. Alagaratnam J, Sharma TK, Lim CS, Fleck BW. A survey of visual impairment using WHO/PBL methodology. *Eye* 2002;**16**:557-61.

209. Hall DM, Hall SM. Early detection of visual defects in infancy. *BMJ* 1988;**296**:823-4.

210. Lambert SR. Management of monocular congenital cataracts. *Eye* 1999;**13**:474-9.

211. Simon JW, Mehta N, Simmons ST. Glaucoma after pediatric lensectomy/vitrectomy. *Ophthalmology* 1991;**98**:670-4.

212. Brady KM, Atkinson CS, Kilty LA, Hiles DA. Glaucoma after cataract extraction and posterior chamber lens implantation in children. *J Cataract Refract Surg* 1997;**23**:669.

213. Miyahara S, Amino K, Tanihara H. Glaucoma secondary to pars plana lensectomy for congenital cataract. *Graefes Arch Clin Exp Ophthalmol* 2002;**240**:176-9.

214. Wallace DK, Plager DA. Corneal diameter in childhood aphakic glaucoma. *J Pediatr Ophthalmol Strabismus* 1996;**33**:230-40.

215. Walton DS. Pediatric aphakic glaucoma: a study of 65 patients. *Tr Am Ophth Soc* 1995;**93**:403-13.

216. Watts P, Abdolell M, Levin A. Complications in infants undergoing surgery for congenital cataract in the first 12 weeks of life: Is early surgery better? *J AAPOS* 2003;7:81-5.

217. Fielder AR, Irwin M, Auld R, Cocker KD, Jones HS, Molesley MJ. Compliance in amblyopia therapy:objective monitoring of occlusion. *Br J Ophthalmol* 1995;**79**:585-9.

218. Loudon SE, Polling JR, Simonsz HJ. A peliminary repot about the relation between visual acuity increase and compliance in patching therapy for amblyopia. *Strabismus* 2002;**10**:79-82.

219. Stewart-Brown S, Haslum M, Butler N. Educational attainment of 10 year old children with treated and untreated visual defects. *Developmental Medicine and Child Neurology* 1985;**27**:504-13.

220. Choong YF, Lukman H, Martin S, Laws DE. Childhood amblyopia treatment:psychosocial implications for patients and primary carers. *Eye* 2004;**18**:369-75.

221. Searle A, Norman P, Harrad R, Vedhara K. Psychosocial and clinical determinants of compliance with occlusion therapy for amblyopic children. *Eye* 2002;**16**:150-5.

222. Levartovsky S, Oliver M, Gottesman N, Shimshoni M. Factors affecting long term results of successfully treated amblyopia:initial visual acuity and type of amblyopia. *Br J Ophthalmol* 1995;**79**:225-8.

223. Gothwal VK, Lovie-Kitchin JE, Nutheti R. The development of the LV Prasad-Functional Vision Questionnaire: a measure of functional vision

performance of visually impaired children. *Invest Ophthalmol Vis Sci* 2003;**44**:4131-9.

224. Eiser C, Vance YH, Horne B, Glaser A, Galvin H. The value of the PEDSQLTM in assessing quality of life in survivors of childhood cancer. *Child Care Health Dev* 2002;**29**:95-102.

225. Varni JW, Seid M, Knight TS, Burwinkle T, Brown J, Szer IS. The PedsQL in paediatric rheumatology. *Arthritis and Rheumatism* 2002;**46**:714-25.

226. Varni JW, Burwinkle T, Katz ER, Meeske K, Dickinson P. The PedsQL™ in pediatric cancer. *Cancer* 2002;**94**:2090-106.

227. Eiser C, Morse R. Quality of Life measures in chronic diseases of childhood. *Health Technology Assessment* 2001;**5**.

9. ABBREVIATIONS

CL	Contact lenses
HRQOL	Health related quality of life
IOL	Intraocular lens implantation
ODM	Occlusion Dose Monitor
PCO	Posterior capsular opacity
PMMA	Polymethyl methacrylate (type of plastic)
QOL	Quality of life
VA	Visual acuity
VRQOL	Vision related quality of life
WHO	World Health Organisation

10.1 Appendix 1

Child and parental parallel PedsQL 4.0™ Questionnaires

The questionnaire is suitable for children from 2 to 18. Included is the questionnaire suitable for 8-12 year olds

ID# _____

Date: _____

PedsQL™

Pediatric Quality of Life Inventory

Version 4.0

CHILD REPORT (ages **8-12**)

DIRECTIONS

On the following page is a list of things that might be a problem for you. Please tell us **how much of a problem** each one has been for you during the **past ONE month** by circling:

0 if it is **never** a problem
1 if it is **almost never** a problem
2 if it is **sometimes** a problem
3 if it is **often** a problem
4 if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

*In the past **ONE month**, how much of a **problem** has this been for you ...*

ABOUT MY HEALTH AND ACTIVITIES *(problems with...)*	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard for me to walk more than one block	0	1	2	3	4
2. It is hard for me to run	0	1	2	3	4
3. It is hard for me to do sports activity or exercise	0	1	2	3	4
4. It is hard for me to lift something heavy	0	1	2	3	4
5. It is hard for me to take a bath or shower by myself	0	1	2	3	4
6. It is hard for me to do chores around the house	0	1	2	3	4
7. I hurt or ache	0	1	2	3	4
8. I have low energy	0	1	2	3	4

ABOUT MY FEELINGS *(problems with...)*	Never	Almost Never	Some-times	Often	Almost Always
1. I feel afraid or scared	0	1	2	3	4
2. I feel sad or blue	0	1	2	3	4
3. I feel angry	0	1	2	3	4
4. I have trouble sleeping	0	1	2	3	4
5. I worry about what will happen to me	0	1	2	3	4

HOW I GET ALONG WITH OTHERS *(problems with...)*	Never	Almost Never	Some-times	Often	Almost Always
1. I have trouble getting along with other kids	0	1	2	3	4
2. Other kids do not want to be my friend	0	1	2	3	4
3. Other kids tease me	0	1	2	3	4
4. I cannot do things that other kids my age can do	0	1	2	3	4
5. It is hard to keep up when I play with other kids	0	1	2	3	4

ABOUT SCHOOL *(problems with...)*	Never	Almost Never	Some-times	Often	Almost Always
1. It is hard to pay attention in class	0	1	2	3	4
2. I forget things	0	1	2	3	4
3. I have trouble keeping up with my schoolwork	0	1	2	3	4
4. I miss school because of not feeling well	0	1	2	3	4
5. I miss school to go to the doctor or hospital	0	1	2	3	4

ID# _____

Date: _____

PedsQL™

Pediatric Quality of Life Inventory

Version 4.0

PARENT REPORT for **CHILDREN** (ages **8-12**)

DIRECTIONS

On the following page is a list of things that might be a problem for **your child**. Please tell us **how much of a problem** each one has been for **your child** during the **past ONE month** by circling:

0 if it is **never** a problem
1 if it is **almost never** a problem
2 if it is **sometimes** a problem
3 if it is **often** a problem
4 if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question, please ask for help.

*the past **ONE month,** how much of a **problem** has your child had with ...*

PHYSICAL FUNCTIONING ***(problems with...)***	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Walking more than one block	0	1	2	3	4
2. Running	0	1	2	3	4
3. Participating in sports activity or exercise	0	1	2	3	4
4. Lifting something heavy	0	1	2	3	4
5. Taking a bath or shower by him or herself	0	1	2	3	4
6. Doing chores around the house	0	1	2	3	4
7. Having hurts or aches	0	1	2	3	4
8. Low energy level	0	1	2	3	4

EMOTIONAL FUNCTIONING ***(problems with...)***	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Feeling afraid or scared	0	1	2	3	4
2. Feeling sad or blue	0	1	2	3	4
3. Feeling angry	0	1	2	3	4
4. Trouble sleeping	0	1	2	3	4
5. Worrying about what will happen to him or her	0	1	2	3	4

SOCIAL FUNCTIONING ***(problems with...)***	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Getting along with other children	0	1	2	3	4
2. Other kids not wanting to be his or her friend	0	1	2	3	4
3. Getting teased by other children	0	1	2	3	4
4. Not able to do things that other children his or her age can do	0	1	2	3	4
5. Keeping up when playing with other children	0	1	2	3	4

SCHOOL FUNCTIONING ***(problems with...)***	**Never**	**Almost Never**	**Some-times**	**Often**	**Almost Always**
1. Paying attention in class	0	1	2	3	4
2. Forgetting things	0	1	2	3	4
3. Keeping up with schoolwork	0	1	2	3	4
4. Missing school because of not feeling well	0	1	2	3	4
5. Missing school to go to the doctor or hospital	0	1	2	3	4

10.2 Appendix 2

Questionnaire to investigate the national outcomes of congenital cataract

Outcomes in the British Congenital/Infantile Cataract Study Cohort in collaboration with the *British Congenital Cataract Interest Group (BCCIG).*

PART 1
Please complete ALL sections

Question 1: Classification Of Cataract

Question II: Additional Non-Ophthalmic Disorders

Question III: Initial Management Confirmation

Question IV: Most Recent Ophthalmic Assessment

Question V: Treatment For Amblyopia

Question VI: Blind Or Partially Sighted Certification

Question VII: Education And Special Needs

PLEASE RETURN THIS FORM IN THE PREPAID ENVELOPE TO:

Melanie Chak, Clinical Research Fellow, Centre for Paediatric Epidemiology and Biostatistics,
Institute of Child Health
Tel:
Fax:
Email:

Outcomes in the British Congenital/Infantile Cataract Study Cohort in collaboration with the *British Congenital Cataract Interest Group (BCCIG).*

FIRST 3 LETTERS OF PATIENT'S SURNAME:

PATIENT'S HOSPITAL NO.:

PATIENT'S DATE OF BIRTH:

Is this patient still under your care?

☐ Yes
☐ No ☐ Patient died
☐ Patient lost to follow up
☐ Patient referred/transferred to: **Consultant Name and Address:**

Date of referral

DATE OF THE LAST RECORDED DATA

PLEASE REPORT INFORMATION AFTER THIS DATE IN THIS FORM

QUESTION I. CLASSIFICATION OF CATARACT

Please update/amend current information on laterality and classification of cataract

a. Previously recorded laterality of cataract ☐Unilateral right ☐Unilateral left ☐Bilateral

b. Has the laterality of the cataract changed ☐No, unchanged ☐Yes, now bilateral, specify date

c. Previously reported underlying or associated causes of cataract.

d. Have underlying or associated cause(s) of the cataract changed?

☐No, unchanged ☐Yes, specify new cause(s):

e. Date of revised diagnosis

QUESTION II. ADDITIONAL NON OPHTHALMIC DISORDERS

Please update/amend current information on non-ophthalmological disorders

a. Previously reported additional non-ophthalmological disorders

b. Any subsequent additional non-ophthalmological disorders? ☐No, ☐Yes, specify

c. Any additional non-ophthalmological impairments? ☐No ☐Yes, specify

☐Hearing ☐Mobility ☐Global developmental delay ☐Speech/Language ☐Learning/Intellectual ☐Other, specify

QUESTION III. INITIAL MANAGEMANT CONFIRMATION

Please update/amend info on INITIAL management. Part 2 relates to subsequent management.

	Right Eye	**Left Eye**
a.Initial reported management (please amend as necessary)	☐Conservative treatment ☐Lens aspiration with vitrectomy ☐Lens aspiration no vitrectomy ☐Phacoemulsification ☐Lensectomy – vitrectomy: ☐Pars plana ☐Cornea ☐ Other, specify ☐**IOL** ☐**No IOL**	☐Conservative treatment ☐Lens aspiration with vitrectomy ☐Lens aspiration no vitrectomy ☐Phacoemulsification ☐Lensectomy – vitrectomy: ☐Pars plana ☐Cornea ☐ Other, specify ☐**IOL** ☐**No IOL**
b.Complications reported to date (please amend as necessary)	☐No ☐Yes,specify ☐Anaesthetic related, ☐periop ☐postop ☐Lens dislocation ☐periop ☐postop ☐Iris trauma/prolapse ☐periop ☐postop ☐Posterior capsular opacity ☐Excessive postop uveitis ☐Wound leak ☐periop ☐postop ☐Hyphaema ☐periop ☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop ☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage ☐Other, specify	☐No ☐Yes,specify ☐Anaesthetic related, ☐periop ☐postop ☐Lens dislocation ☐periop ☐postop ☐Iris trauma/prolapse ☐periop ☐postop ☐Posterior capsular opacity ☐Excessive postop uveitis ☐Wound leak ☐periop ☐postop ☐Hyphaema ☐periop ☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop ☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage ☐Other, specify

QUESTION IV MOST RECENT OPHTHALMIC ASSESSMENT

DATE of examination	

a. **FIXATION** ☐ NOT APPLICABLE / RECORDED

	Right eye	Left eye
CENTRAL	☐Yes ☐No	☐Yes ☐No
STEADY	☐Yes ☐No	☐Yes ☐No
MAINTAINED	☐Yes ☐No	☐Yes ☐No

b. **VISUAL ACUITY**

DISTANCE: ☐ NOT APPLICABLE / RECORDED

Method:	☐LogMAR system ☐Sonksen Silver Acuity System ☐Snellen optotypes ☐Kay's pictures ☐Sheridan Gardner singles ☐ Other (specify) :
Test distance *(if applicable)*:	☐**without correction** ☐**with correction** ☐spectacles ☐contact lens
Visual acuity ***(actual** measurement and with Snellen equivalent if possible)* **Right** **Left** **Both eyes together**	

NEAR: ☐ NOT APPLICABLE / NOT RECORDED

Method (specify) :	**Test distance**(specify):	☐**without correction** ☐**with correction** ☐spectacles ☐contact lens
Right	**Left**	**Both eyes together**

c. **STEREOPSIS:** ☐ NOT APPLICABLE / RECORDED

Method (specify): ☐Titmus fly ☐Frisby ☐Randot ☐Lang ☐Other, specify

Stereopsis measurement

d. **RETINOSCOPY FINDINGS:** ☐ NOT APPLICABLE / RECORDED

RETINOSCOPY	
Right eye	
Left eye	
Cycloplegic agent ☐YES ☐ NO	**Working distance** ☐50 cms ☐75cms ☐Other, specify

e. **CLINICAL EXAMINATION**

Extra-ocular movements:	☐Normal	☐Abnormal (specify):
Strabismus	☐No	☐Left / ☐Right / ☐ Alt. Angle: ☐Constant ☐Intermittent ☐Esotropia ☐Exotropia ☐Vertical ☐ Other :
Nystagmus	☐No	☐Latent / ☐ Manifest ☐Horizontal ☐Vertical ☐Combined ☐Pendular ☐Jerk ☐Rotary ☐Combined ☐ Other :

Right eye		Left eye
☐Normal ☐Abnormal, specify	**cornea** incl clarity & horizontal diameter (mm)	☐Normal ☐Abnormal, specify
☐Normal ☐Abnormal, specify	**anterior chamber**	☐Normal ☐Abnormal, specify
☐Normal ☐Abnormal, specify	**iris / pupil** including pupil reaction	☐Normal ☐Abnormal, specify
☐Phakic, normal ☐Aphakic ☐Pseudophakic ☐Phakic, specify cataract type	**lens morphology**	☐Phakic, normal ☐Aphakic ☐Pseudophakic ☐Phakic, specify cataract type
	vitreous	
	IOP (mmHg) instrument used	
	fundus examination	
☐Normal ☐Abnormal, specify	disc	☐Normal ☐Abnormal, specify
☐Normal ☐Abnormal, specify	macula	☐Normal ☐Abnormal, specify
☐Normal ☐Abnormal, specify	vessels	☐Normal ☐Abnormal, specify
☐Normal ☐Abnormal, specify	peripheral retina	☐Normal ☐Abnormal, specify

f. OPHTHALMIC INVESTIGATIONS ***Please record relevant/recent results***

ELECTRORETINOGRAM: ☐ NOT APPLICABLE / RECORDED

STIMULUS: ☐**FLASH** ☐**PATTERN**

Right	☐ **Not done**	☐**Normal**	☐**Abnormal***specify*
Left	☐ **Not done**	☐**Normal**	☐**Abnormal***specify*
Binocular	☐ **Not done**	☐**Normal**	☐**Abnormal***specify*

VISUAL EVOKED POTENTIAL: ☐ NOT APPLICABLE / RECORDED

STIMULUS: ☐**FLASH** ☐**PATTERN** ☐**SWEEP**

Right	☐ **Not done**	☐**Normal**	☐**Abnormal***specify*
Left	☐ **Not done**	☐**Normal**	☐**Abnormal***specify*
Binocular	☐ **Not done**	☐**Normal**	☐**Abnormal***specify*

OTHER INVESTIGATIONS

Test and Findings:

QUESTION V. TREATMENT FOR AMBLYOPIA

a. Has this child ever undergone: occlusion? ☐Yes ☐No **or penalisation ?** ☐Yes ☐No

b. Was occlusion/penalisation commenced
☐Pre-operatively ☐Post-operatively ☐Other, specify

c. Percentage of intended regime achieved
☐100% ☐75% ☐50% ☐less than 50%

d. Concordance record: ☐ No ☐ Yes: checked with ☐Diary ☐Log book ☐Other

e. Concordance ☐ Full ☐ Greater than 50% ☐Less than 50% ☐ None

f. Main reason for poor concordance
☐Uncooperative child ☐Parental reluctance ☐Other, specify

g. Was occlusion/penalisation abandoned?
☐No ☐Yes, specify ☐Parental decision ☐Clinician decision (poor visual prognosis) ☐Other, specify

QUESTION VI. BLIND OR PARTIALY SIGHTED CERTIFICATION

a. Has this child been certified? ☐No: ☐Ineligible ☐Parents declined ☐Other, specify

☐Yes: ☐Blind ☐Partially sighted

Date certified

QUESTION VII. EDUCATION AND SPECIAL NEEDS: ☐Known ☐ Not known

a. School attended by subject?
☐Mainstream, no additional teaching ☐Special needs school
☐Mainstream, with additional teaching ☐Other, specify

b. For any child>16 years old *(please tick all that apply):*
☐Further/higher education:☐ AS levels☐ A levels☐City and Guilds/Diplomas☐University Degree☐Other, specify

☐Employed, specify occupation

c. Does the child receive any ADDITIONAL educational input outside of school? *(please tick all that apply)*
☐ From education services, specify

☐ From charitable organisations, specify

☐ Other, specify

d. Visual disability the PRIMARY reason that additional teaching or special needs schooling was sought? ☐Yes☐No
If no, specify primary reason

e. Any other special needs support?
☐No ☐Yes, specify

NAME OF PERSON WHO COMPLETED THE FORM

PLEASE USE THIS BOX FOR ANY COMMENTS

THANK YOU FOR COMPLETING PART 1 OF THE FORM

PLEASE RETURN THIS FORM IN THE PREPAID ENVELOPE TO:

Melanie Chak, Clinical Research Fellow
Centre for Paediatric Epidemiology and Biostatistics
Institute of Child Health

Outcomes in the British Congenital/Infantile Cataract Study Cohort in collaboration with the *British Congenital Cataract Interest Group (BCCIG).*

PART 2

Only CERTAIN sections apply to a given child. The prompts⇒ at the beginning of each section will guide you.

Question VIII: Surgery For Cataract

Question IX: IOL Implantation

Question X: Secondary Membrane Formation Or Posterior Capsular Opacity

Question XI: Glaucoma

Question XII: Squint surgery

Question XIII: Post-operative Uveitis

Question XIV: Retinal Detachment

Question XV: Endophthalmitis

Question XVI: Other Complications

PLEASE RETURN THIS FORM IN THE PREPAID ENVELOPE TO:
Melanie Chak, Clinical Research Fellow, Centre for Paediatric Epidemiology and Biostatistics, Institute of Child Health

Outcomes in the British Congenital/Infantile Cataract Study Cohort in collaboration with the *British Congenital Cataract Interest Group (BCCIG).*

FIRST 3 LETTERS OF PATIENT'S SURNAME:

PATIENT'S HOSPITAL NO.:

PATIENT'S DATE OF BIRTH:

Is this patient still under your care?

☐ Yes
☐ No ☐ Patient died
☐ Patient lost to follow up
☐ Patient referred/transferred to: **Consultant Name and Address:**

Date of referral

DATE OF THE LAST RECORDED DATA

PLEASE REPORT INFORMATION AFTER THIS DATE IN THIS FORM

⇒☐**Child had cataract surgery after initial conservative treatment** PLEASE FILL IN QUESTION VIII, PAGE 3

⇒☐**Child had cataract develop in a previously normal eye and had surgery** PLEASE FILL IN QUESTION VIII, PAGE3

⇒☐**Child had cataract surgery initially and HAD a secondary IOL** PLEASE GO TO QUESTION IX, PAGE 4

⇒☐**Child had cataract surgery initially and NO secondary IOL** PLEASE GO TO QUESTION X, PAGE 4

⇒☐**Child had cataract surgery and primary IOL initially** PLEASE GO TO QUESTION X, PAGE 4

⇒☐**Child NEVER had cataract surgery:** PLEASE GO TO QUESTION XI, PAGE 5

QUESTION VIII. SURGERY FOR CATARACT

	RIGHT EYE	LEFT EYE
a. Did cataract develop in a previously normal eye?	☐No ☐Yes	☐No ☐Yes
b. Was the cataract removed following initial conservative management?	☐No ☐Yes ☐Visual loss/amblyopia ☐Cosmetic appearance ☐Other, specify	☐No ☐Yes ☐Visual loss/amblyopia ☐Cosmetic appearance ☐Other, specify
c. Date of operation		
d. Surgical procedure performed (IOL discussed overleaf)	☐Lens aspiration with vitrectomy ☐Lens aspiration no vitrectomy ☐Phacoemulsification ☐Lensectomy – vitrectomy: ☐Pars plana ☐Cornea ☐ Other, specify	☐Lens aspiration with vitrectomy ☐Lens aspiration no vitrectomy ☐Phacoemulsification ☐ Lensectomy – vitrectomy: ☐Pars plana ☐Cornea ☐ Other, specify
e. Primary capsulotomy/vitrectomy?	☐No ☐Yes, specify ☐YAG capsulotomy ☐Anterior capsulorrhexis/otomy ☐Posterior capsulorrhexis ☐Vitrectomy	☐No ☐Yes, specify ☐YAG capsulotomy ☐Anterior capsulorrhexis/otomy ☐Posterior capsulorrhexis ☐Vitrectomy
	☐By hand ☐Mechanical ☐Unknown	☐By hand ☐Mechanical ☐Unknown
Complications of cataract surgery?	☐No ☐Yes, specify ☐Anaesthetic related, ☐periop ☐postop ☐Lens dislocation ☐periop ☐postop ☐Iris trauma/prolapse ☐periop ☐postop ☐Wound leak ☐periop ☐postop ☐Hyphaema ☐periop ☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop ☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage ☐Other, specify	☐No ☐Yes, specify ☐Anaesthetic related, ☐periop ☐postop ☐Lens dislocation ☐periop ☐postop ☐Iris trauma/prolapse ☐periop ☐postop ☐Wound leak ☐periop ☐postop ☐Hyphaema ☐periop ☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop ☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage ☐Other, specify

THANK YOU, NOW PLEASE PROCEED TO QUESTION IX, PAGE 4

QUESTION IX. IOL IMPLANTATION

⇒1 IOL implant (either eye)
☐Yes **PLEASE CONTINUE WITH THIS QUESTION** ☐No **PLEASE PROCEED TO QUESTION X, BELOW**

	Right eye	Left eye
	Date of IOL placement: ☐Primary ☐Secondary	Date of IOL placement: ☐Primary ☐Secondary
a. State where the IOL was placed	☐Bag ☐Sulcus ☐Ant chamber ☐Unknown ☐Other, specify	☐Bag ☐Sulcus ☐Ant chamber ☐Unknown ☐Other, specify
b. IOL Manufacturer		
c. IOL Material	☐PMMA only ☐PMMA heparin coated ☐Acrylic ☐Silicone ☐Other,specify	☐PMMA only ☐PMMA heparin coated ☐Acrylic ☐Silicone ☐Other,specify
d. IOL design	☐Foldable ☐One piece ☐Non-foldable ☐Three piece ☐Other,specify	☐Foldable ☐One piece ☐Non-foldable ☐Three piece ☐Other,specify
e. IOL Power		
f. Formula used	☐Hoffer Q ☐Holladay ☐SRKII ☐SRKT ☐Other, specify	☐Hoffer Q ☐Holladay ☐SRKII ☐SRKT ☐Other, specify
g. Final intended refraction, after eye settled postop		

THANK YOU, NOW PLEASE PROCEED TO QUESTION X, BELOW.

QUESTION X. SECONDARY MEMBRANE FORMATION OR POSTERIOR CAPSULAR OPACITY

⇒1 **Did either posterior capsular opacity or secondary membrane formation occur?**
☐Yes **PLEASE CONTINUE WITH THIS QUESTION.** ☐No **PLEASE PROCEED TO QUESTION XI.**

	Right eye	Left eye
	☐ Posterior capsular opacity ☐ Secondary membrane formation	☐ Posterior capsular opacity ☐ Secondary membrane formation
a. Date of treatment		
b. Treatment	☐YAG capsulotomy: ☐LA ☐GA ☐Surgical capsulotomy: ☐pars plana ☐limbal	☐YAG capsulotomy: ☐LA ☐GA ☐Surgical capsulotomy: ☐pars plana ☐limbal
c. Number of procedures needed?	☐ One ☐>1, specify	☐ One ☐>1, specify
d. Clinically significant complications of laser/surgery for posterior capsular opacification or secondary membrane formation?	☐No ☐Yes,specify ☐Anaesthetic related, ☐periop ☐postop ☐Lens pitting ☐Lens dislocation ☐periop ☐postop ☐Iris trauma/prolapse ☐periop ☐postop ☐Excessive postop uveitis ☐Wound leak ☐periop ☐postop ☐Hyphaema ☐periop ☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop ☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage ☐Other, specify	☐No ☐Yes,specify ☐Anaesthetic related, ☐periop ☐postop ☐Lens pitting ☐Lens dislocation ☐periop ☐postop ☐Iris trauma/prolapse ☐periop ☐postop ☐Excessive postop uveitis ☐Wound leak ☐periop ☐postop ☐Hyphaema ☐periop ☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop ☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage ☐Other, specify

THANK YOU, NOW PLEASE PROCEED TO QUESTION XI, PAGE 5.

QUESTION XI. GLAUCOMA

⇒1 **Was glaucoma ever diagnosed in this child?**
☐Yes **PLEASE CONTINUE WITH THIS QUESTION.** ☐ No **PLEASE PROCEED TO QUESTION XII, PAGE 6.**

	Right eye	**Left eye**
a. Diagnosis date		
b. Diagnostic criteria used (tick all that apply)	☐Raised IOP ☐Optic disc appearance ☐Myopic Shift ☐Increasing corneal diameter ☐Corneal Haze ☐Field analysis ☐Humphrey ☐Goldman ☐Estermann ☐Other, specify	☐Raised IOP ☐Optic disc appearance ☐Myopic Shift ☐Increasing corneal diameter ☐Corneal Haze ☐Field analysis ☐Humphrey ☐Goldman ☐Estermann ☐Other, specify
c. Glaucoma type	**Primary glaucoma** ☐ Primary congenital glaucoma ☐ Anterior segment dysgenesis, specify ☐ Systemic syndrome, specify **Secondary glaucoma** ☐ Postoperative open angle glaucoma ☐Postoperative closed angle glaucoma with pupil block ☐Postoperative closed angle glaucoma without pupil block ☐Steroid induced ☐Trauma ☐**Other glaucomas**, specify	**Primary glaucoma** ☐ Primary congenital glaucoma ☐ Anterior segment dysgenesis, specify ☐ Systemic syndrome, specify **Secondary glaucoma** ☐ Postoperative open angle glaucoma ☐Postoperative closed angle glaucoma with pupil block ☐Postoperative closed angle glaucoma without pupil block ☐Steroid induced ☐Trauma ☐**Other glaucomas**, specify
d. Medical treatment (please tick all that apply)	☐Betoptic ☐Trusopt ☐Cosopt ☐Betaxolol ☐Teoptic ☐Xalatan ☐Timoptol ☐Alphagan ☐Xalacom ☐Pilocarpine ☐Other, specify	☐Betoptic ☐Trusopt ☐Cosopt ☐Betaxolol ☐Teoptic ☐Xalatan ☐Timoptol ☐Alphagan ☐Xalacom ☐Pilocarpine ☐Other, specify
e. Surgical Treatment?	☐No ☐Yes, specify ☐Goniotomy ☐Trabeculectomy alone ☐Trabeculectomy & 5FU ☐Trabeculectomy &MMC ☐Drainage tubes ☐Other, specify	☐No ☐Yes, specify ☐Goniotomy ☐Trabeculectomy alone ☐Trabeculectomy & 5FU ☐Trabeculectomy &MMC ☐Drainage tubes ☐Other, specify
f. Date of surgical treatment		
g. Laser Treatment?	☐No ☐Yes, specify ☐Cycloablation ☐Trabeculoplasty ☐Iridotomy ☐Other, specify	☐No ☐Yes, specify ☐Cycloablation ☐Trabeculoplasty ☐Iridotomy ☐Other, specify
h. Date of laser treatment		

i. Additional procedures undertaken for glaucoma?	☐No ☐Yes, specify: ☐Repeat of original procedure No. performed__ ☐Repeat of original procedure with addition of antimetabolite No. performed __ ☐Needling of bleb No. performed__ ☐Needling of bleb with 5FU No. performed__ ☐Further laser No. performed__ ☐Other, specify	☐No ☐Yes, specify: ☐Repeat of original procedure No. performed__ ☐Repeat of original procedure with addition of antimetabolite No. performed __ ☐Needling of bleb No. performed__ ☐Needling of bleb with 5FU No. performed__ ☐Further laser No. performed__ ☐Other, specify
j. Clinically significant complications of surgery or laser treatment for glaucoma (Please tick all that apply)	☐No ☐Yes, specify: ☐Anaesthetic related, ☐periop☐postop ☐Iris trauma/prolapse ☐periop☐postop ☐Excessive postop uveitis ☐Wound leak ☐periop☐postop ☐Hyphaema ☐periop☐postop ☐Excessive filtration ☐periop☐postop ☐Malignant glaucoma ☐Pupil block ☐Non functioning bleb ☐periop☐postop ☐Blebitis ☐periop☐postop ☐Retinal detachment ☐Endophthalmitis ☐Other, specify	☐No ☐Yes, specify: ☐Anaesthetic related, ☐periop☐postop ☐Iris trauma/prolapse ☐periop☐postop ☐Excessive postop uveitis ☐Wound leak ☐periop☐postop ☐Hyphaema ☐periop☐postop ☐Excessive filtration ☐periop☐postop ☐Malignant glaucoma ☐Pupil block ☐Non functioning bleb ☐periop☐postop ☐Blebitis ☐periop☐postop ☐Retinal detachment ☐Endophthalmitis ☐Other, specify

THANK YOU, NOW PLEASE PROCEED TO QUESTION XII, BELOW.

QUESTION XII. SQUINT SURGERY

⇒1 Has the child ever had surgery to correct a squint?

☐Yes **PLEASE CONTINUE WITH THIS QUESTION** ☐No **PLEASE GO TO QUESTION XIII, PAGE 7**

	Right eye	**Left Eye**
a. Date of procedure		
b. Amblyopia associated strabismus	☐Yes ☐No	☐Yes ☐No
c. Surgical procedure performed *(please tick all those that apply)*	☐Lateral rectus recession ☐Medial rectus recession ☐Lateral rectus resection ☐Medial rectus resection ☐Inferior oblique myomectomy ☐Inferior oblique recession ☐Faden procedure ☐Other, specify	☐Lateral rectus recession ☐Medial rectus recession ☐Lateral rectus resection ☐Medial rectus resection ☐Inferior oblique myomectomy ☐Inferior oblique recession ☐Faden procedure ☐Other, specify
d. Surgical complications of squint surgery?	☐Infection ☐periop ☐postop ☐Excessive haemorrhage ☐periop ☐postop ☐Overcorrection ☐periop ☐postop ☐Undercorrection ☐periop ☐postop ☐Other, specify	☐Infection ☐periop ☐postop ☐Excessive haemorrhage ☐periop ☐postop ☐Overcorrection ☐periop ☐postop ☐Undercorrection ☐periop ☐postop ☐Other, specify
e. Further operations necessary?	☐No ☐Yes, specify	☐No ☐Yes, specify

THANK YOU, NOW PLEASE PROCEED TO QUESTION XIII, PAGE 7.

QUESTION XIII. INCREASED POST-OPERATIVE- UVEITIS

⇒1 **Has the child ever had clinically significant postoperative uveitis?**
☐Yes **PLEASE CONTINUE WITH THIS QUESTION** ☐No **PLEASE GO TO QUESTION XIV, BELOW.**

	Right eye	Left eye
a. The normal regime for postop cataract surgery?		
b. Additional treatment necessary? (Please tick all that apply)	☐increased topical meds ☐oral steroids ☐orbital floor injections ☐subconj injections Specify meds used ☐Surgery, specify	☐increased topical meds ☐oral steroids ☐orbital floor injections ☐subconj injections Specify meds used ☐Surgery, specify
c. Any possible associations with uveitis (Please tick all that apply)	☐ Non-concordance with medication ☐ Afro-Carribean descent ☐ Systemic inflamm disease, specify ☐Other, specify	☐ Non-concordance with medication ☐ Afro-Carribean descent ☐ Systemic inflamm disease, specify ☐Other, specify

THANK YOU, NOW PLEASE PROCEED TO QUESTION XIV, BELOW.

QUESTION XIV. RETINAL DETACHMENT

⇒1 **Has the child ever had a retinal detachment?**
☐Yes **PLEASE CONTINUE WITH THIS QUESTION** ☐No **PLEASE GO TO QUESTION XV, PAGE 8.**

	Right eye	Left Eye
a. Was retinal detachment a direct complication of cataract surgery?	☐Yes ☐No If no, specify aetiology if known	☐Yes ☐No If no, specify aetiology if known
b. Surgical intervention? (Please tick all that apply)	☐No ☐Yes, specify ☐Vitrectomy ☐Gas ☐Oil ☐Plomb ☐External drainage ☐Cryotherapy ☐Laser ☐Other, specify	☐No ☐Yes, specify ☐Vitrectomy ☐Gas ☐Oil ☐Plomb ☐External drainage ☐Cryotherapy ☐Laser ☐Other, specify
c. Clinically significant complications of retinal detachment surgery?	☐No ☐Yes,specify ☐Anaesthetic related,☐periop☐postop ☐Excessive postop uveitis ☐Wound leak ☐periop☐postop ☐Hyphaema ☐periop☐postop ☐High postop IOP ☐Secondary cataract ☐Vitreous loss/ ☐periop☐postop strand to wound ☐Further retinal detachment ☐Cystoid mac oedema ☐Endophthalmitis ☐Other, specify	☐No ☐Yes,specify ☐Anaesthetic related,☐periop☐postop ☐Excessive postop uveitis ☐Wound leak ☐periop☐postop ☐Hyphaema ☐periop☐postop ☐High postop IOP ☐Secondary cataract ☐Vitreous loss/ ☐periop☐postop strand to wound ☐Further retinal detachment ☐Cystoid mac oedema ☐Endophthalmitis ☐Other, specify
d. Further surgery necessary?	☐No ☐Yes, specify: Date of surgery Surgical intervention	☐No ☐Yes, specify: Date of surgery Surgical intervention

THANK YOU, NOW PLEASE PROCEED TO QUESTION XV, PAGE 8.

QUESTION XV. ENDOPHTHALMITIS

⇒1 Has the child ever had endophthalmitis?
☐Yes **PLEASE CONTINUE WITH THIS QUESTION** ☐No **PLEASE GO TO QUESTION XVI, BELOW.**

	Right Eye	Left Eye
a. Endophthalmitis associated with cataract surgery?	☐Yes ☐No, specify aetiology	☐Yes ☐No, specify aetiology
b. Date of diagnosis		
c. Diagnosis by	☐Clinical appearance ☐Vitrectomy biopsy ☐Vitreous needle biopsy ☐Anterior chamber tap ☐Other, specify	☐Clinical appearance ☐Vitrectomy biopsy ☐Needle biopsy ☐Anterior chamber tap ☐Other, specify
d. Pathogen isolated	☐No pathogen☐Viral☐Fungal☐Bacterial Specify organism(s)	☐No pathogen☐Viral☐Fungal☐Bacterial Specify organism(s)
e. Treatment	☐Intravitreal antibiotics, specify ☐Topical antibiotics, specify ☐Topical steroids ☐IV antibiotics, specify	☐Intravitreal antibiotics, specify ☐Topical antibiotics, specify ☐Topical steroids ☐IV antibiotics, specify

THANK YOU, NOW PLEASE PROCEED TO QUESTION XVI, BELOW.

QUESTION XVI. OTHER COMPLICATIONS OR SURGICAL PROCEDURES

⇒1 **Has the child had any clinically significant postoperative complication or any other surgical procedure not covered elsewhere in the questionnaire**
☐Yes **PLEASE CONTINUE WITH THIS QUESTION (Copies of this question available at end of form)**
☐No **THANK YOU FOR COMPLETING PART 2 OF THIS FORM Please turn overleaf**

	Right eye	Left eye
a. Surgery or procedure resulting in complication		
b. Complication	☐No ☐Yes,specify ☐Anaesthetic related,☐periop☐postop ☐Lens dislocation ☐periop☐postop ☐Iris trauma/prolapse☐periop☐postop ☐Wound leak ☐periop☐postop ☐Hyphaema ☐periop☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage☐Other, specify	☐No ☐Yes,specify ☐Anaesthetic related,☐periop☐postop ☐Lens dislocation ☐periop☐postop ☐Iris trauma/prolapse☐periop☐postop ☐Wound leak ☐periop☐postop ☐Hyphaema ☐periop☐postop ☐Glaucoma ☐Vitreous loss/ strand to wound ☐periop☐postop ☐Retinal detachment ☐Endophthalmitis ☐Cystoid mac oedema ☐Expulsive haemorrhage☐Other, specify
c. Date of surgery/complication		
d. Treatment required		

NAME OF PERSON WHO COMPLETED THE FORM

PLEASE USE THIS BOX FOR ANY COMMENTS

THANK YOU FOR COMPLETING PART 2 OF THE FORM

PLEASE RETURN THIS FORM IN THE PREPAID ENVELOPE TO:

Melanie Chak, Clinical Research Fellow
Centre for Paediatric Epidemiology and Biostatistics
Institute of Child Health

10.3 Appendix 3

The Contact lens/Patching/Glasses Questionnaire

THE CONTACT LENS/PATCHING/GLASSES QUESTIONNAIRE

With this questionnaire we are hoping to build a picture of how your child has been since starting treatment with **contact lenses, glasses, and/or patching.** We recognise that your child may have had a complicated treatment regime which may have stopped and started and changed over the years. What we are trying to achieve is an **OVERALL** impression of what it has been like for you, your child and your family to have undergone these treatments. We would like to know of any effects (good or bad) which you feel having contact lenses, patching, atropine drops and/or glasses may have had on your child and your family.

- The questionnaire should only take a few minutes to fill in.

- Some questions are open ended for you to write in your own comments. Others give a selection of phrases for you to choose from. Only answer the questions that apply to your child, eg: if your child never wore glasses, miss out the section on glasses.

- The following is an example of the kinds of questions you will be asked. This example also shows what your answer could mean:

How often do questionnaires that you receive provide you with clear instructions on how to use them:

All the time **most of the time** **sometimes** **occasionally** **never**

So if you circled **'All the time'** it means that questionnaires you receive are always clear on their instructions

Or if you circled **'Sometimes'** it means that the questionnaires you receive are clear in what they want you to do some of the time and some of the time they are not clear

Alternatively if you circled **'Never'** it means that the questionnaires you receive are never clear

IF YOU HAVE ANY QUESTIONS PLEASE CONTACT ME AT THE ADDRESS OR TELEPHONE NUMBER BELOW.

Please return the questionnaire in the stamped, addressed envelope provided to:
Dr Melanie Chak
Department of Epidemiology and Ophthalmology
Institute of Child Health

Thank you very much for your time and effort.

Fill in this page of questions if your child *ever wore/is wearing* CONTACT LENSES.

☐My child has worn/is wearing contact lenses **continue with QUESTION 1**
☐My child has never worn contact lenses **turn the page to QUESTION 2:**

QUESTION 1

a) How old was your child when they started wearing contact lenses?

b) Is your child still wearing contact lenses?

Yes/No (Please delete)

c) Did/is your child managing to wear the contact lenses as often as recommended?

Yes/No (Please delete)

d) Does your child wear contact lenses as well as glasses?

Yes/No (Please delete)

e) If the answer to question (1d) was YES, Are there some situations when your child prefers wearing contact lenses rather than glasses?

Yes/No (Please delete)

f) If the question to (1e) was YES, Please give details of the circumstances when your child prefers their contact lenses

g) Overall, do you think your child's behaviour has changed because they are wearing/wore contact lenses?
Yes/No (Please delete)
(If YES, Please give details of change and what you feel may have contributed to it):

QUESTION 1 (CONTINUED)

h) Overall, how has *your child* reacted to wearing the contact lenses? (Please circle)

Very positively	Positively	Neither negatively or positively	Negatively	Very negatively

i) Overall, how have *you* found the experience of your child wearing contact lenses? (Please circle)

Very easy	Easy	Neither difficult nor easy	Difficult	Very difficult

j) Have *you* ever been worried that the contact lenses may be harmful to your child? (Please circle)

Never	Yes, occasionally	Yes, sometimes	Yes, often	Yes, all the time

k) Overall, how has wearing contact lenses affected your child's relationship with you (parent or principal carer)?(Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

l) Overall, how has wearing contact lenses affected your child's relationship with their siblings?(Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

m) Overall, how has wearing contact lenses affected your child's relationships with their friends?(Please circle)

Improved a lot	Improved a little	No effect on relationships	Worsened a little	Worsened a lot

n) Overall, how has wearing contact lenses affected relationships between other family members? ie those which don't involve the child who is wearing the contact lenses? (Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

Fill in this page of questions if your child *ever wore/is wearing* GLASSES.

☐ **My child has worn/is wearing glasses continue with question 2**
☐ **My child has never worn glasses turn the page to question 3**

QUESTION 2

a) How old was your child when they started wearing glasses?

b) Is your child still wearing glasses?

Yes/No (Please delete)

c) Did/is your child managing to wear the glasses as often as recommended?

Yes/No (Please delete)

d) Does your child wear glasses as well as contact lenses?

Yes/No (Please delete)

e) If the answer to question (1d) was YES, Are there some situations when your child prefers wearing glasses rather than contact lenses?

Yes/No (Please delete)

f) If the question to (1e) was YES, Please give details of the circumstances when your child prefers their glasses

g) Overall, do you think your child's behaviour has changed because they are wearing/wore glasses?

Yes/No (Please delete)
(If YES, Please give details of change and what you feel may have contributed to it):

QUESTION 2 (CONTINUED)

h) Overall, how has *your child* reacted to wearing the glasses? (Please circle)

Very positively	Positively	Neither negatively or positively	Negatively	Very negatively

i) Overall, how have *you* found the experience of your child wearing glasses? (Please circle)

Very easy	Easy	Neither difficult nor easy	Difficult	Very difficult

j) Have *you* ever been worried that the glasses may be harmful to your child? (Please circle)

Never	Yes, occasionally	Yes, sometimes	Yes, often	Yes, all the time

k) Overall, how has wearing glasses affected your child's relationship with you (parent or principal carer)?(Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

l) Overall, how has wearing glasses affected your child's relationship with their siblings?(Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

m) Overall, how has wearing glasses affected your child's relationships with their friends?(Please circle)

Improved a lot	Improved a little	No effect on relationships	Worsened a little	Worsened a lot

n) Overall, how has wearing glasses affected relationships between other family members? ie those which don't involve the child who is wearing the glasses? (Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

Fill in this page of questions if your child *ever wore/is wearing* PATCHES.

☐ **My child has worn/is wearing patches continue with question 3**
☐ **My child has never worn contact lenses turn the page to question 4:**

QUESTION 3

a) How old was your child when they started wearing patches?

b) Is your child still wearing patches?

Yes/No (Please delete)

c) Did/is your child managing to wear the patches as often as recommended?

Yes/No (Please delete)

d) Overall, do you think your child's behaviour has changed because they are wearing/wore patches?

Yes/No (Please delete)
If YES, Please give details of change and what you feel may have contributed to it):

QUESTION 3 (CONTINUED)

e) Overall, how has *your child* reacted to wearing the patches? (Please circle)

Very positively	Positively	Neither negatively or positively	Negatively	Very negatively

f) Overall, how have *you* found the experience of your child wearing patches? (Please circle)

Very easy	Easy	Neither difficult nor easy	Difficult	Very difficult

g) Have *you* ever been worried that the patches may be harmful to your child? (Please circle)

Never	Yes, occasionally	Yes, sometimes	Yes, often	Yes, all the time

h) Overall, how has wearing patches affected your child's relationship with you (parent or principal carer)?(Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

i) Overall, how has wearing patches affected your child's relationship with their siblings?(Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

j) Overall, how has wearing patches affected your child's relationships with their friends?(Please circle)

Improved a lot	Improved a little	No effect on relationships	Worsened a little	Worsened a lot

k) Overall, how has wearing patches affected relationships between other family members? ie those which don't involve the child who is wearing the glasses? (Please circle)

Improved a lot	Improved a little	No effect on relationship	Worsened a little	Worsened a lot

l) Has wearing patches affected your child's performance at school? (Please circle)

A lot better than expected	A little better than expected	No change from expected	A little worse than expected	A lot worse than expected

Please use this space to tell us about any other effects having contact lenses, glasses or patching has had on your child and your family?

Please feel free to comment on the questionnaire in the space below

THANK YOU VERY MUCH FOR COMPLETING THIS QUESTIONNAIRE

Please return the questionnaire in the stamped, addressed envelope provided to:

Dr Melanie Chak
Department of Epidemiology and Ophthalmology
Institute of Child Health

10.4 Appendix 4

Histograms of the distributions of visual acuity comparing children aged >5 years and those aged <5 years.

Figure 24: Snellen visual acuity of children with bilateral cataracts by age at last follow up

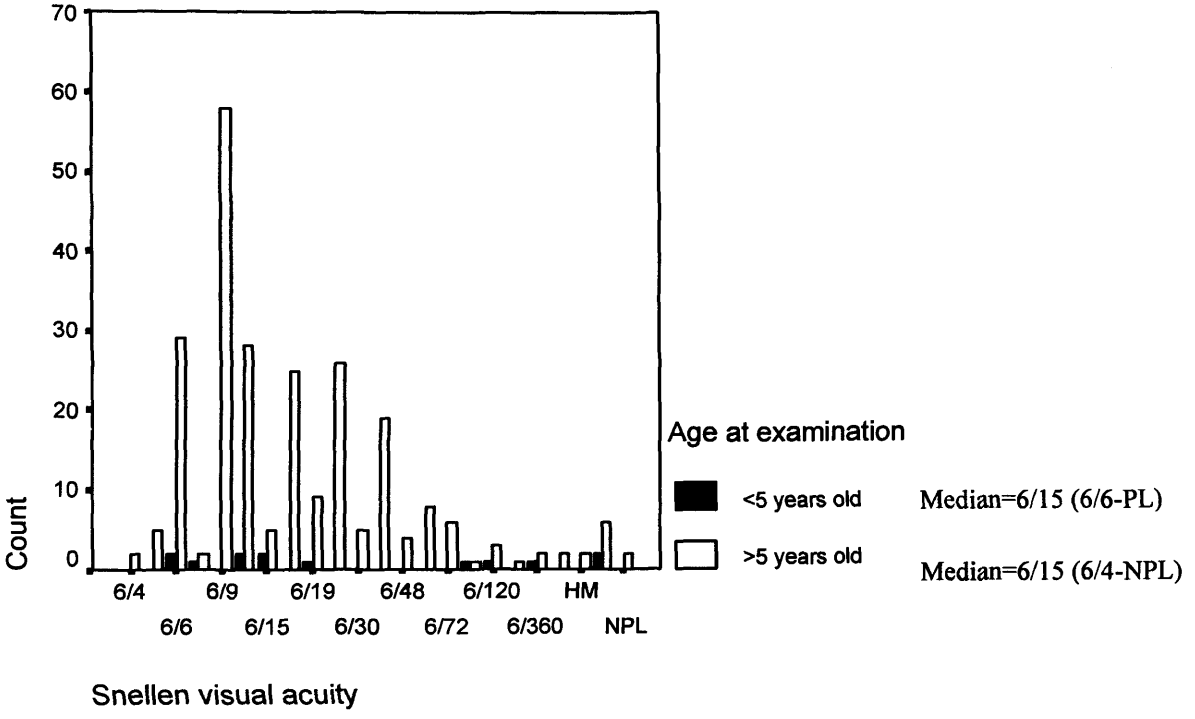


Figure 25: Snellen visual acuity of the eyes with cataract of children with unilateral cataracts by age at last follow up examination

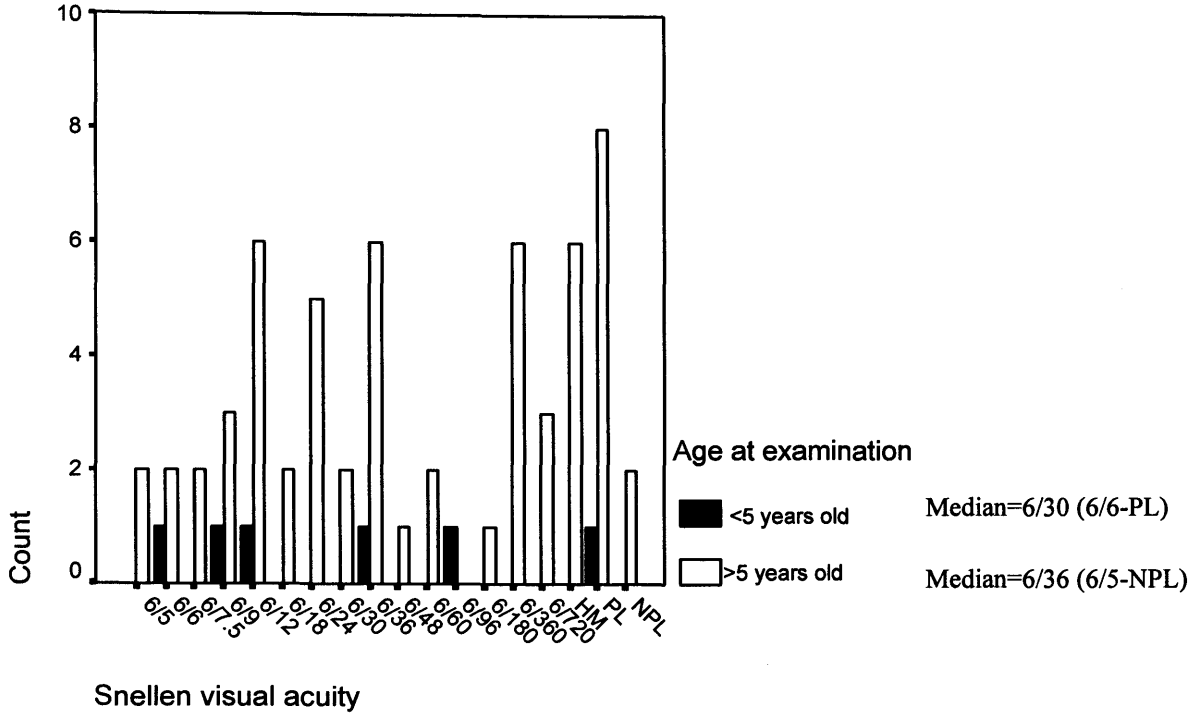
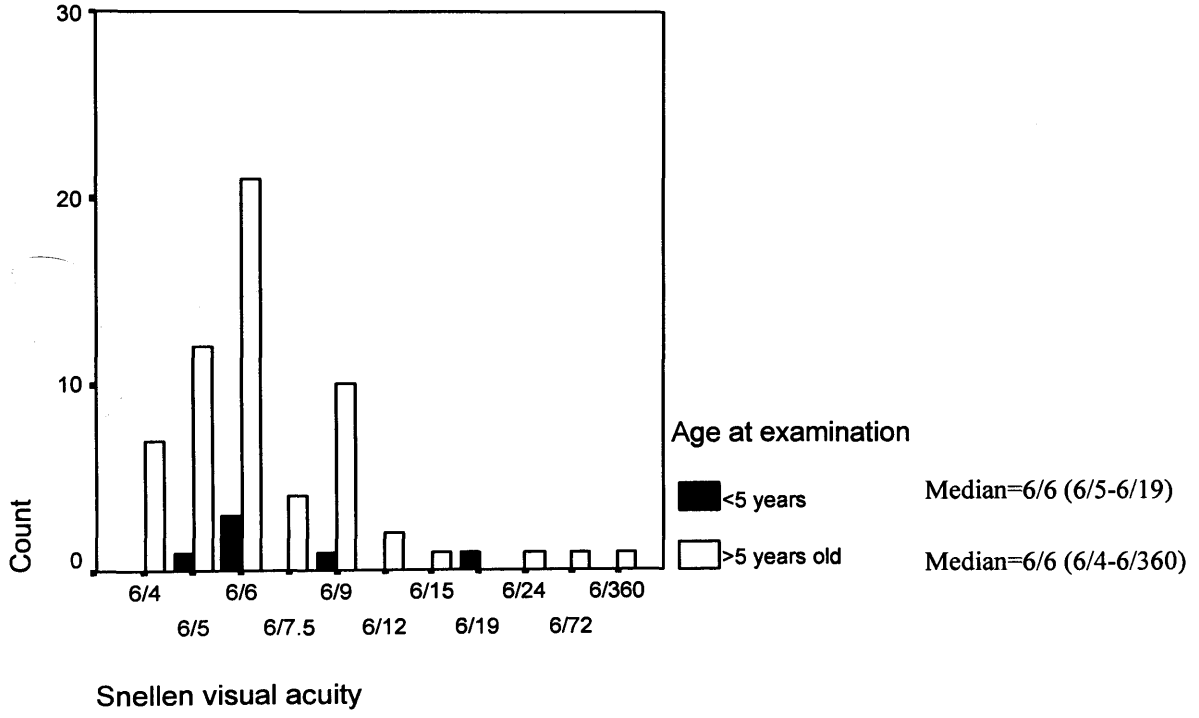


Figure 26: Snellen visual acuity of the eyes without cataract of children with unilateral cataracts separated by age at last follow up examination



10.5 Appendix 5

Factors of interest in relation to the visual acuity of eyes of children aged >5 years old, with bilateral cataracts were assessed univariately as shown in Figures 30 to 42.

Figure 30: Visual acuity at last examination in 236 eyes of children with bilateral cataract by time since detection (months)

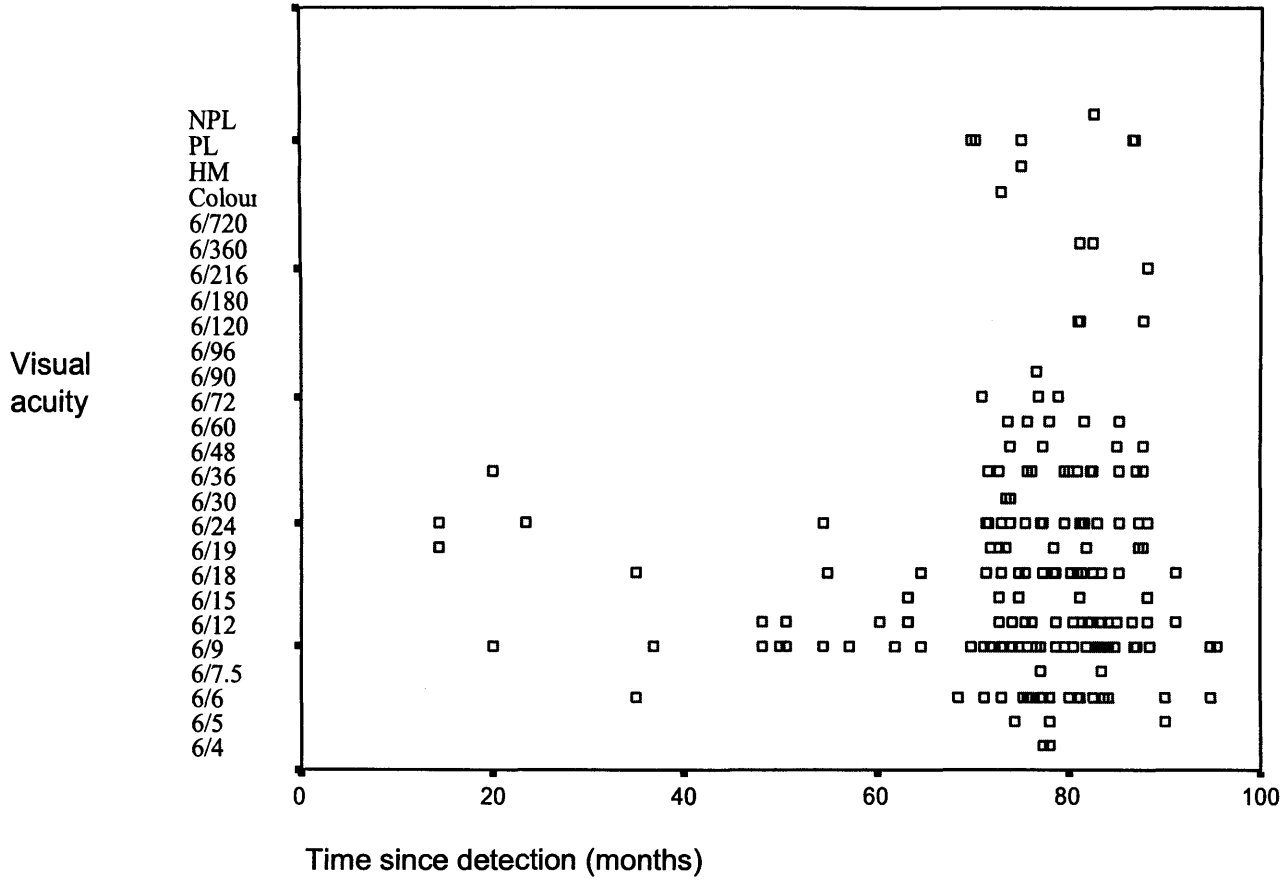


Figure 31: Visual acuity at last examination in 236 eyes of children with bilateral cataract by age at detection (months)

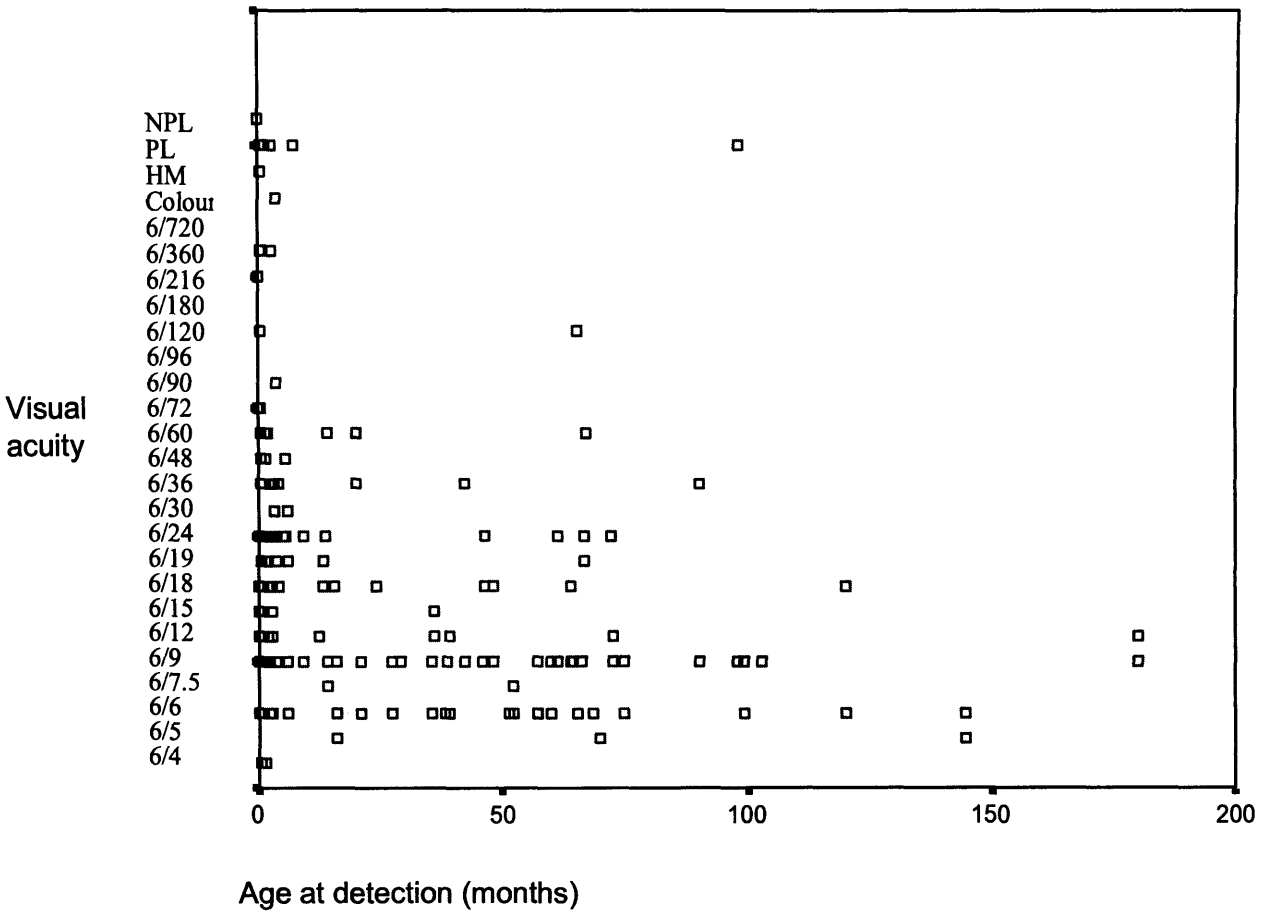


Figure 32: Visual acuity at last examination in 174 eyes of children with bilateral cataract by time since cataract surgery (months)

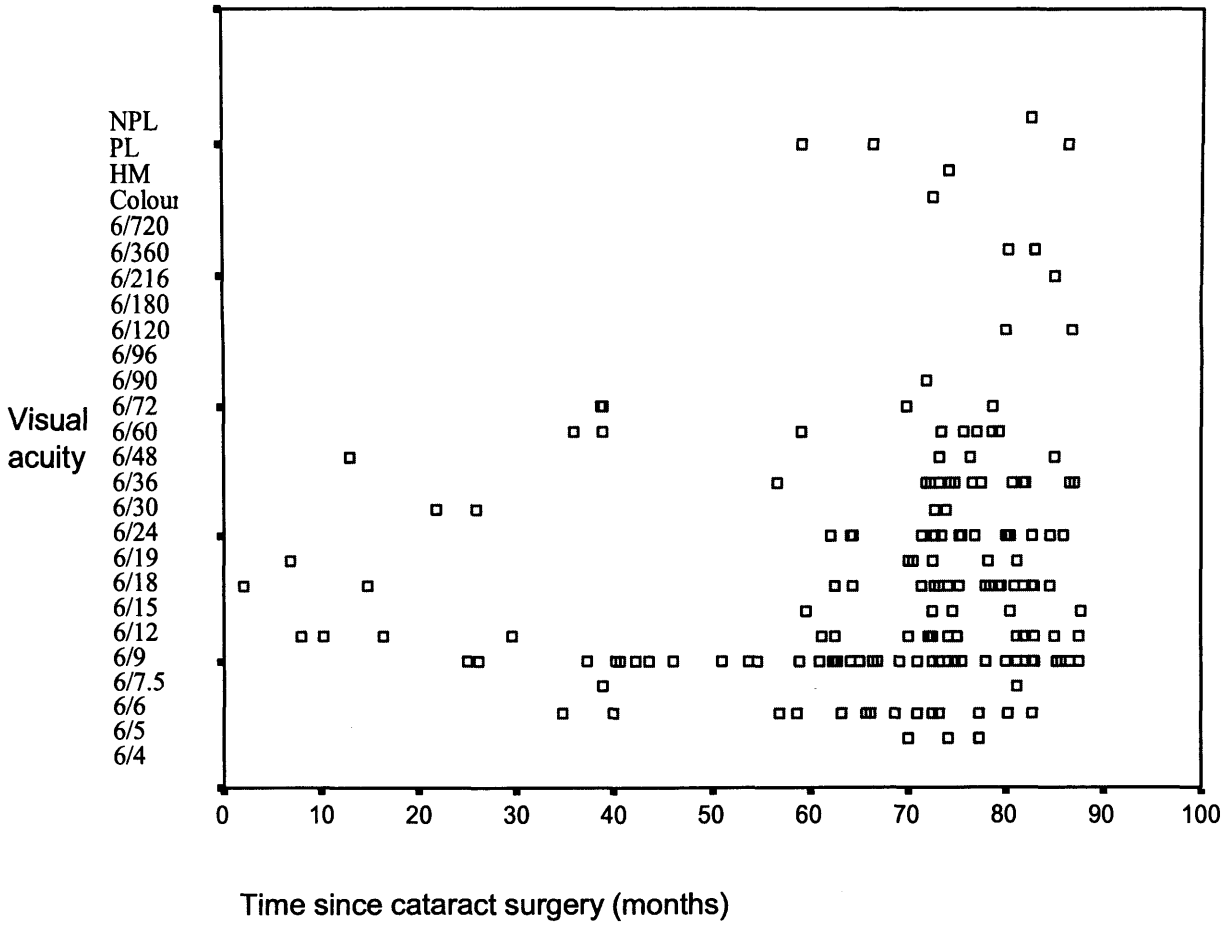


Figure 33: Visual acuity at last examination in 174 eyes of children with bilateral cataract by age at cataract surgery (months)

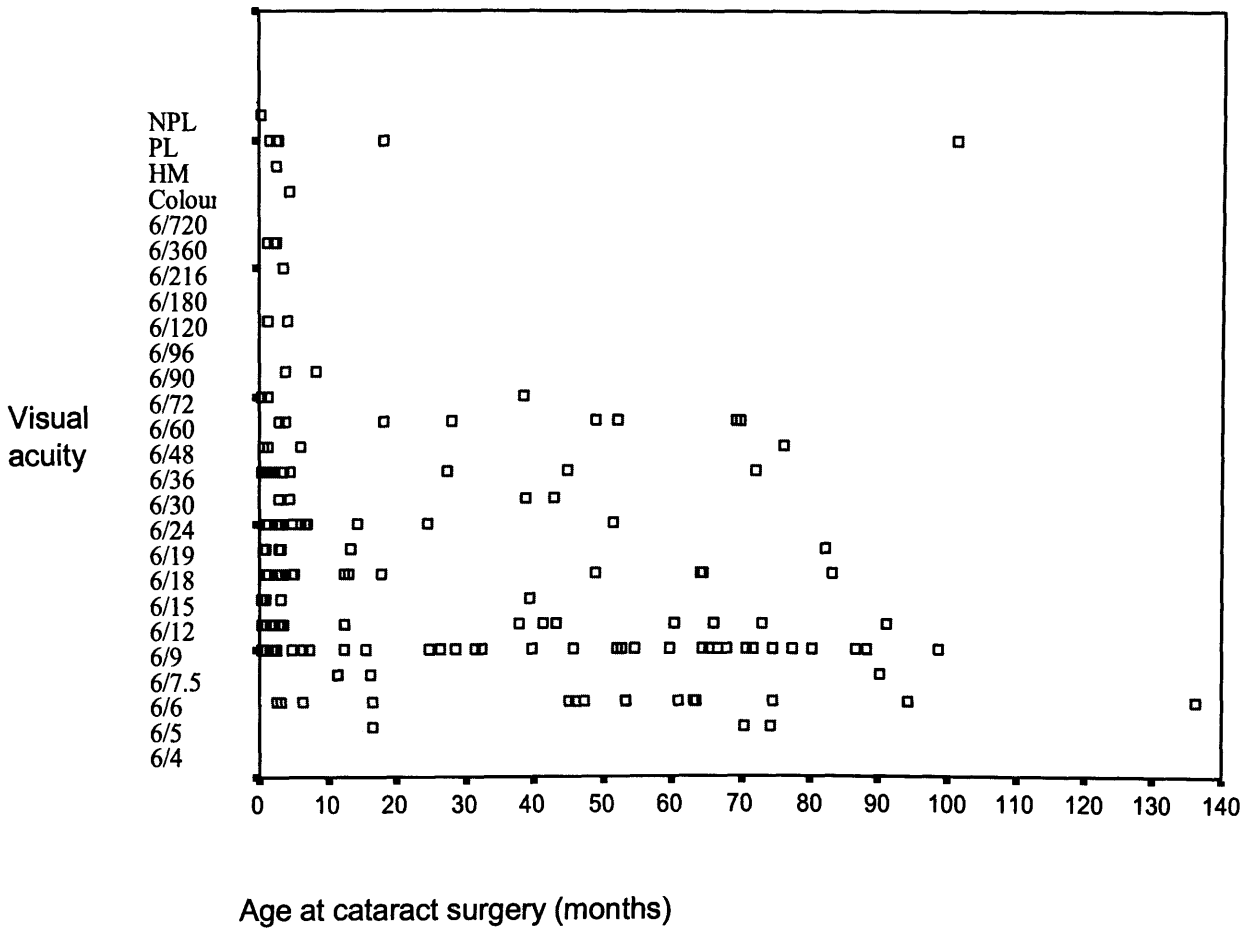


Figure 34: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by final type of correction
(N=178/235)

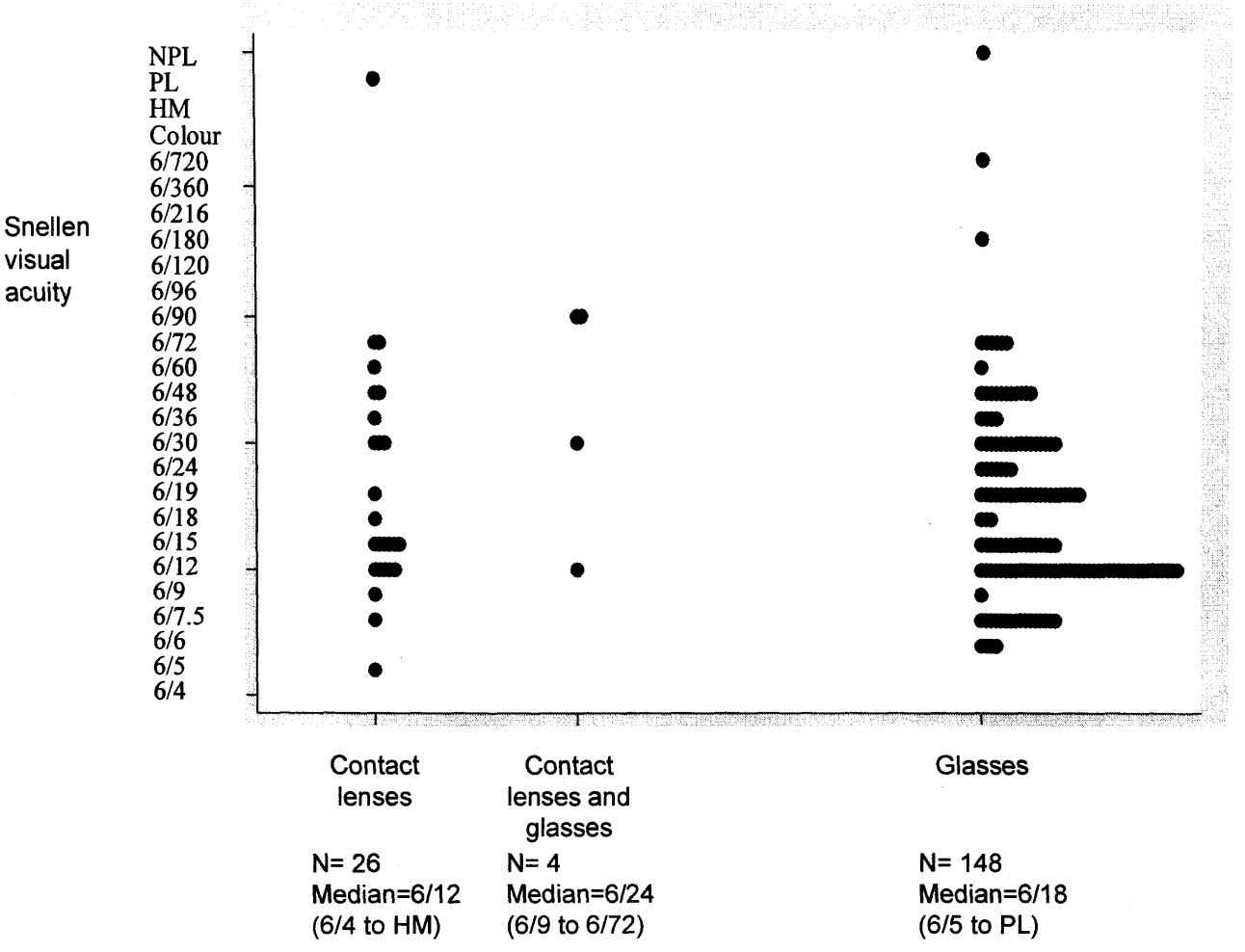


Figure 35: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by type of cataract surgery
(N=174/235)

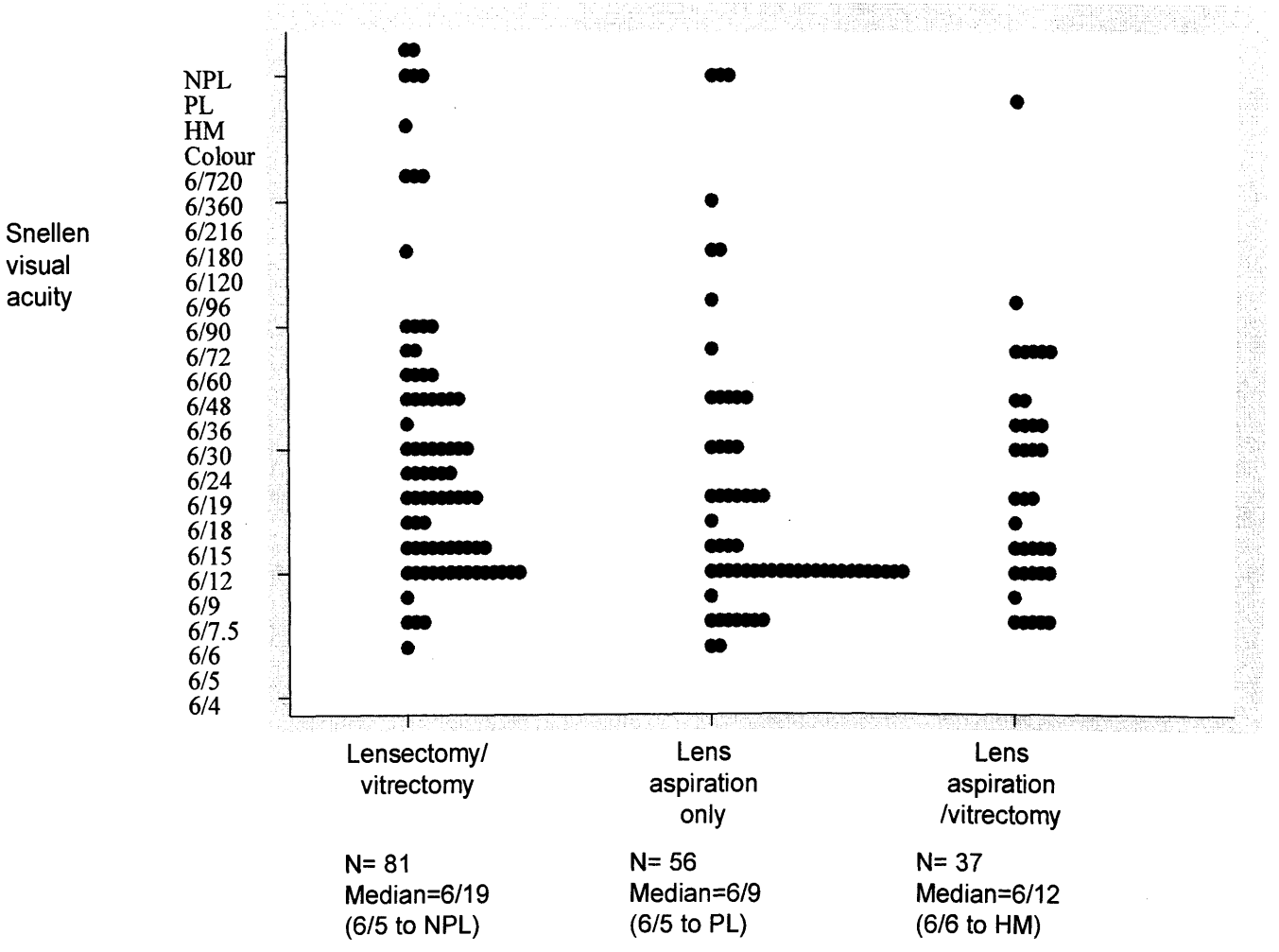


Figure 36: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by primary intraocular lens implantation (N=181/235)

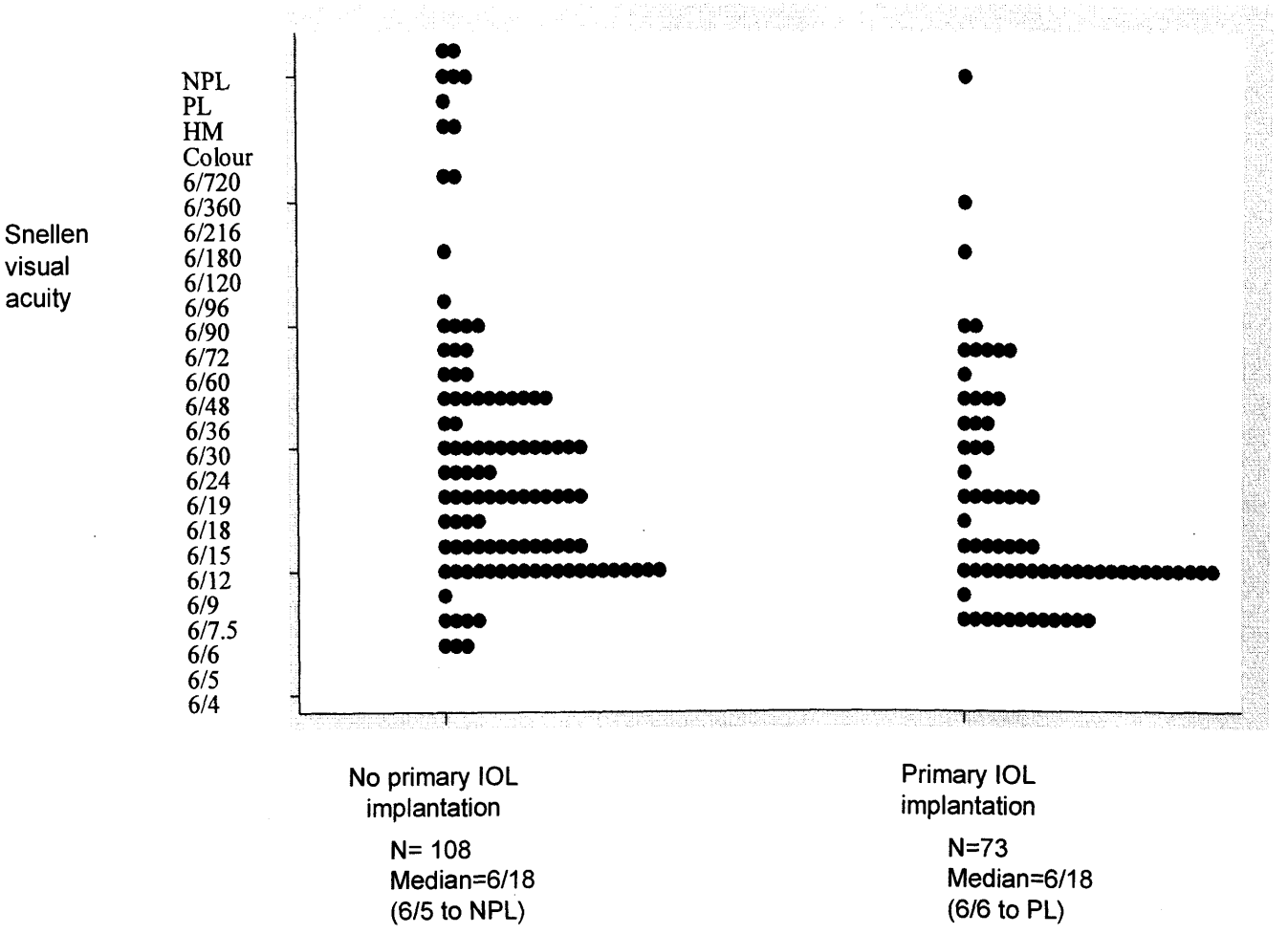


Figure 37: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by concordance with occlusion regime
(N=98/235)

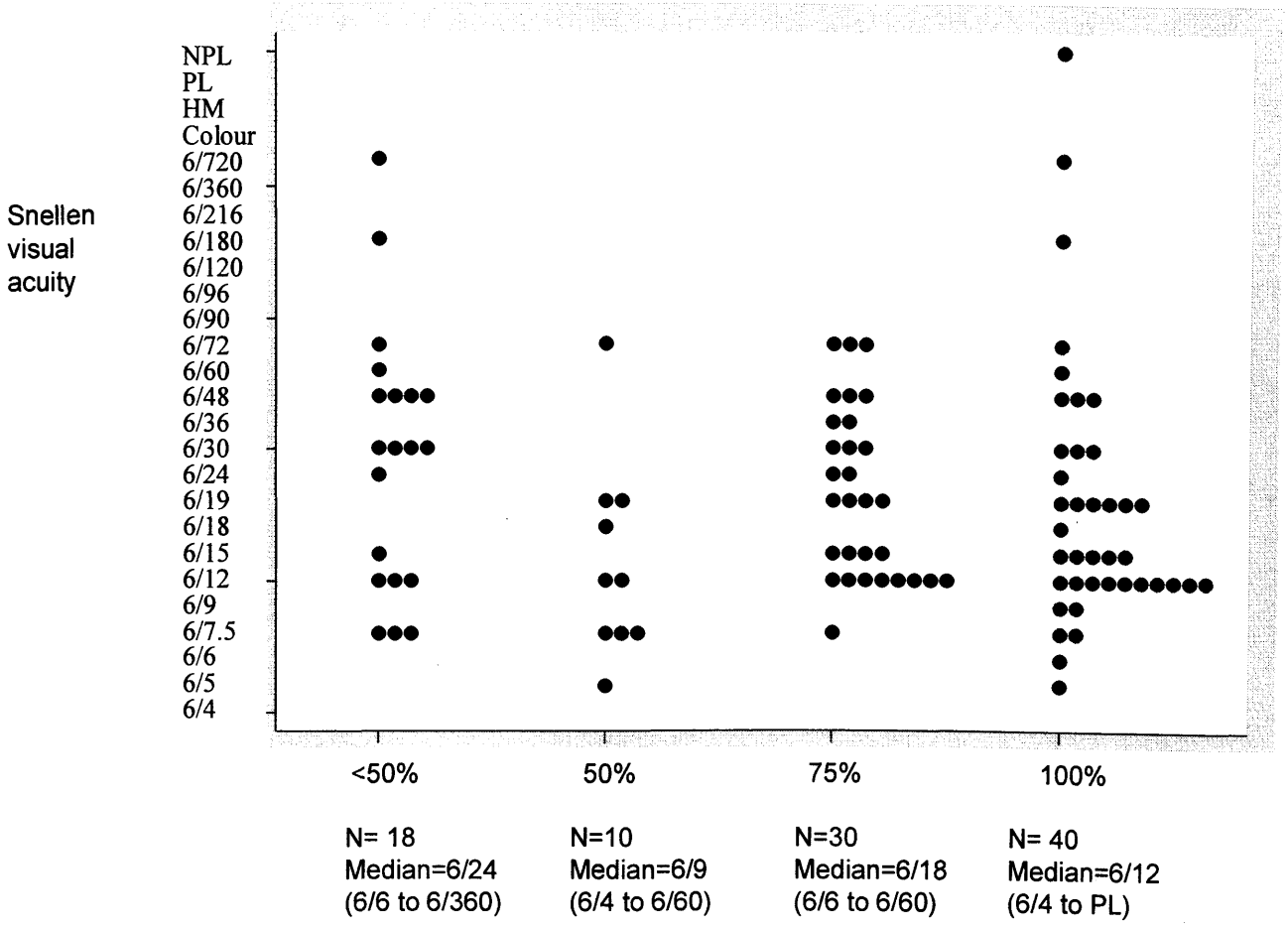


Figure 38: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by aetiological category (N=246/235)

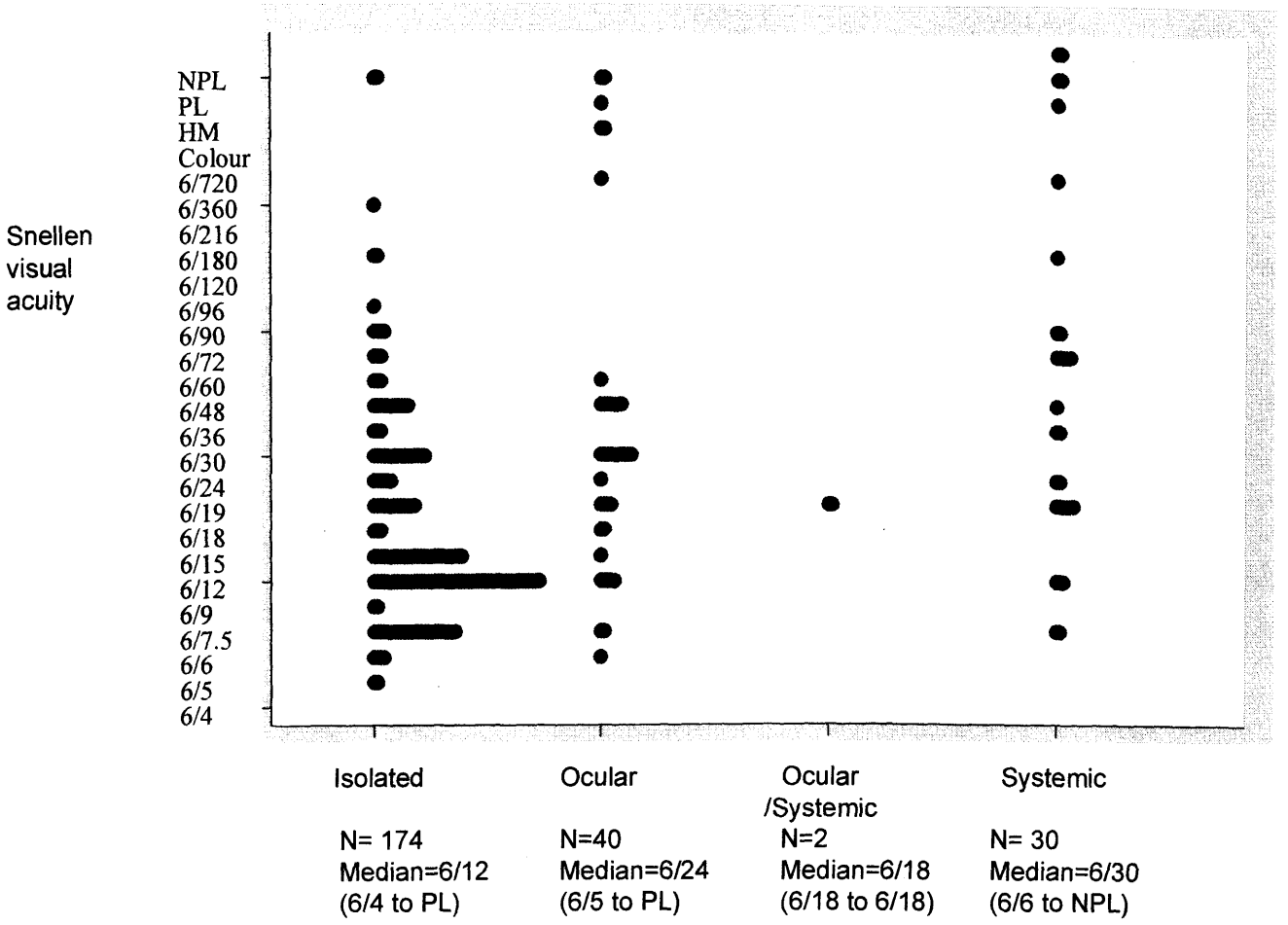


Figure 39: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by presence of other non-ophthalmic medical disorders (N=246/235)

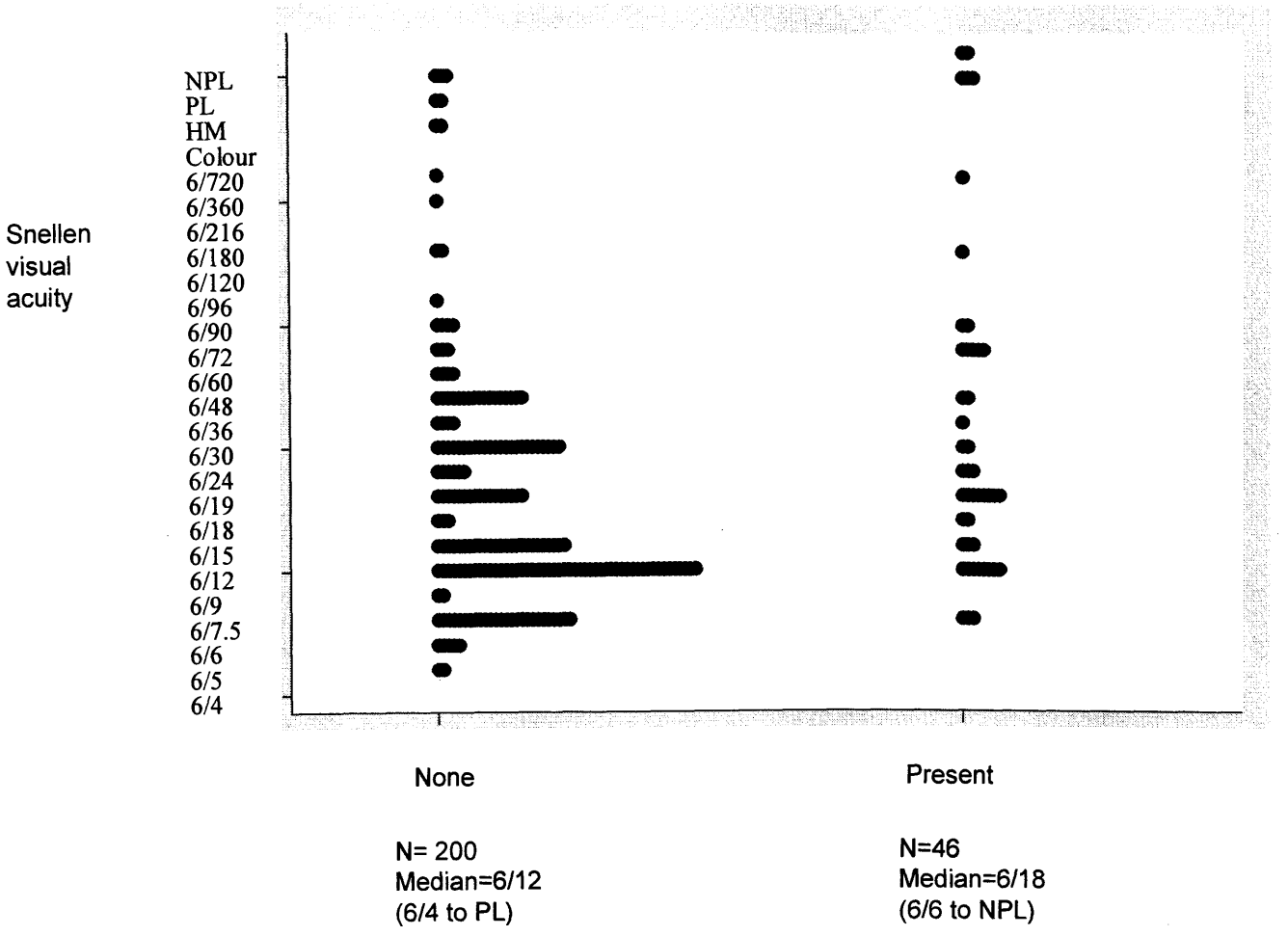


Figure 40: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by severity of cataract at presentation
(N=246/235)

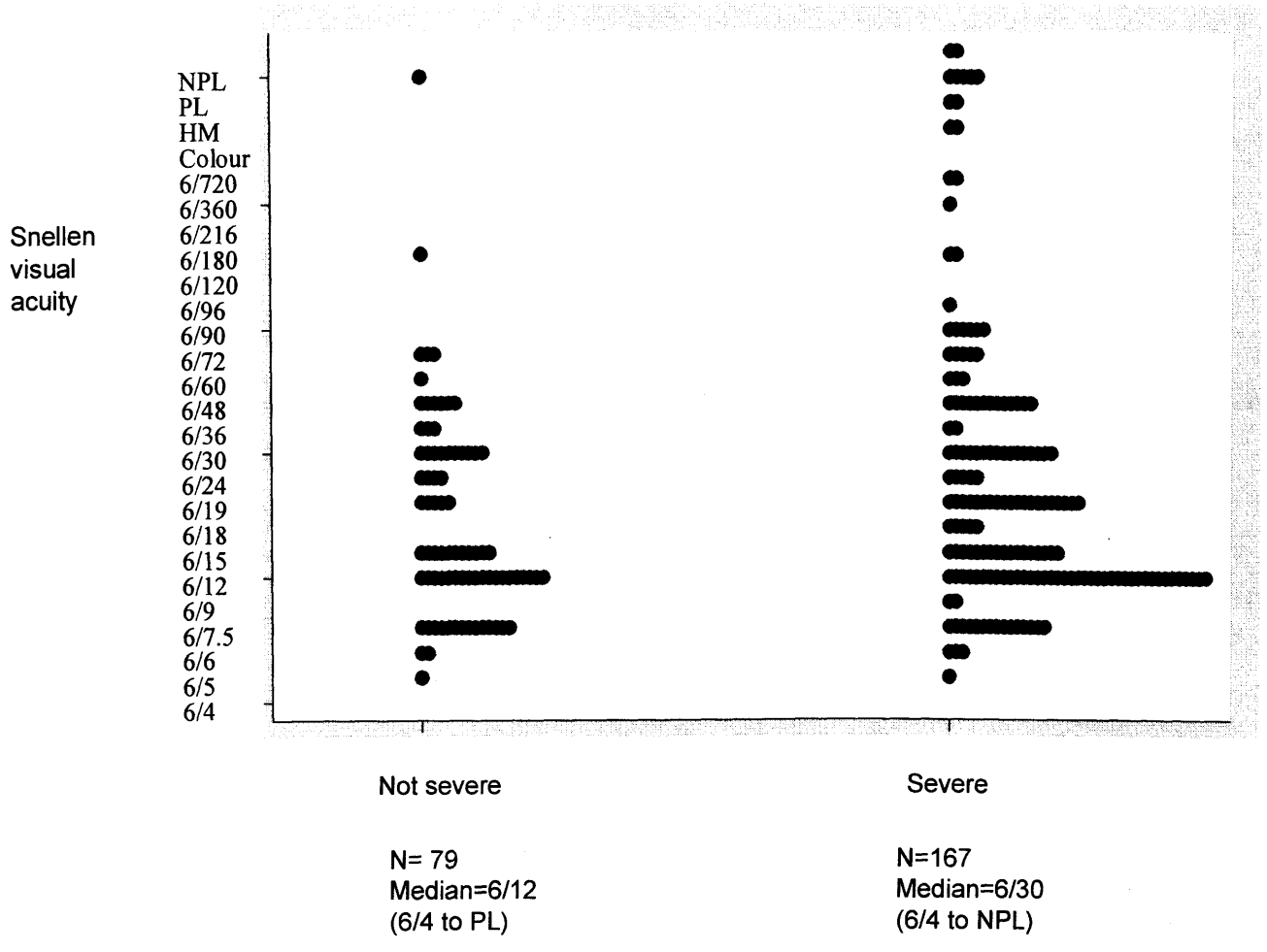


Figure 41: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by occurrence of any postoperative sight threatening complications (N=181/235)

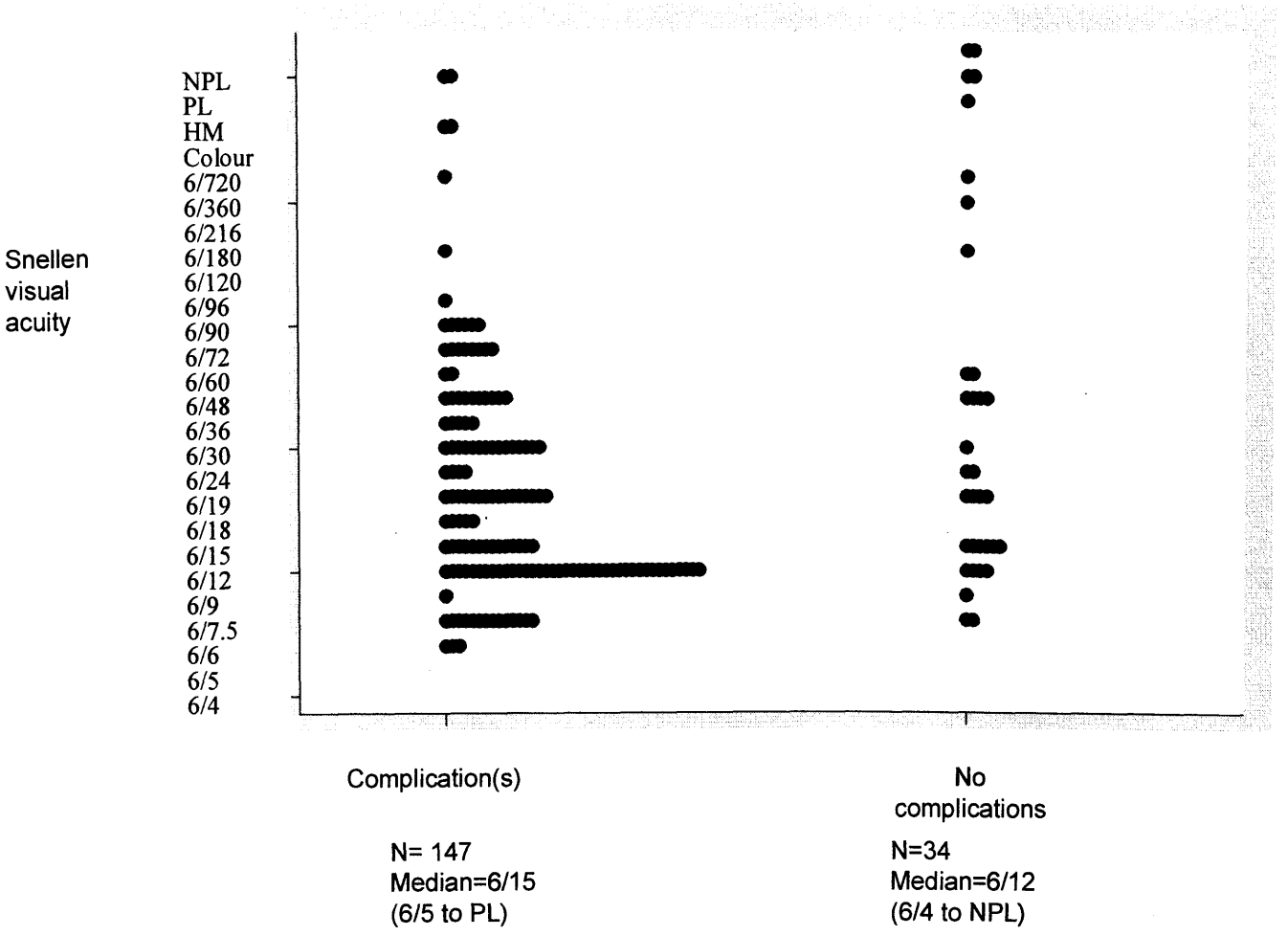
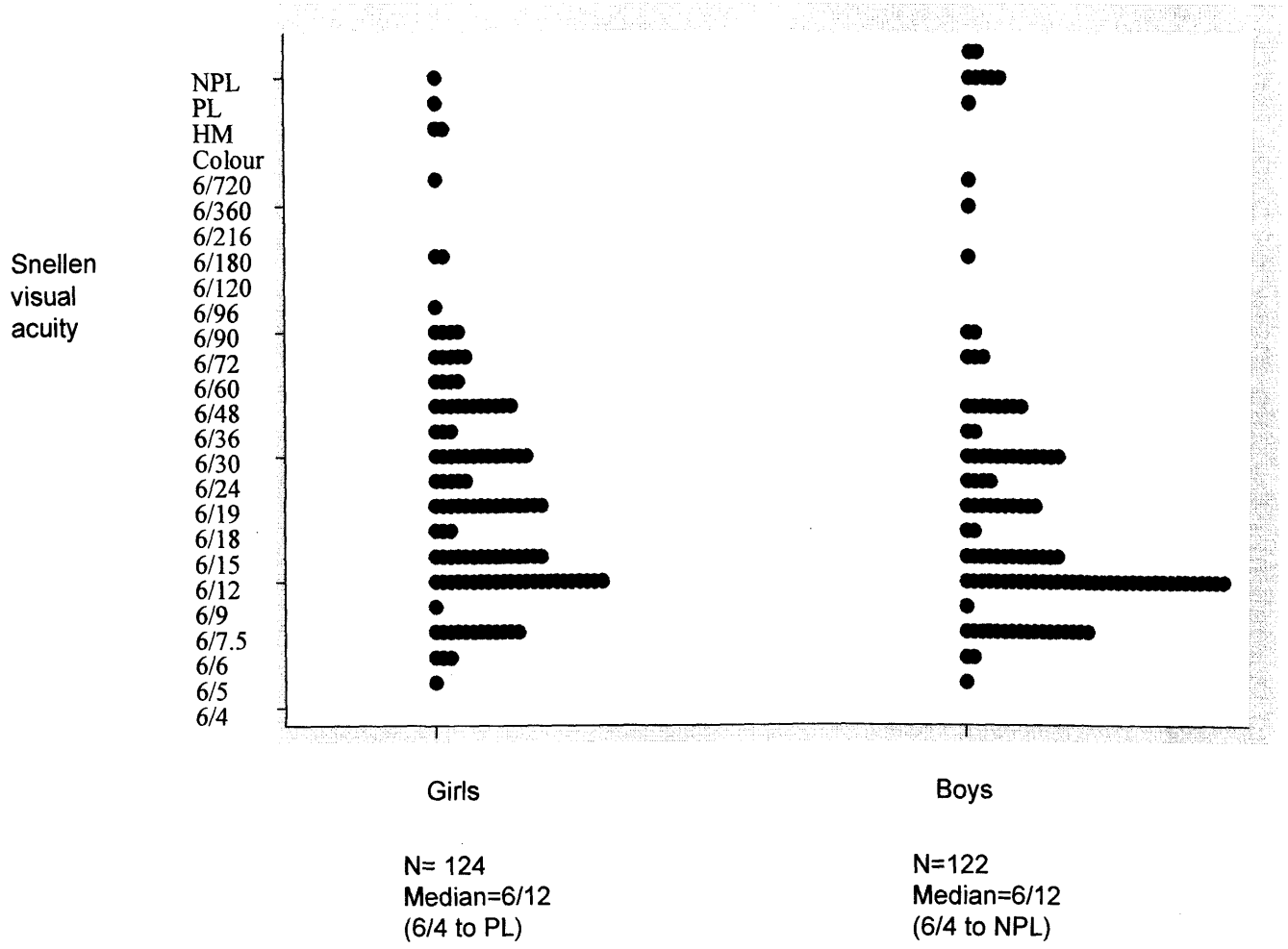


Figure 42: Distribution of visual acuity of eyes of children with bilateral cataract at final examination by gender (N=246/235)



10.6 Appendix 6

Factors of interest in relation to the visual acuity of eyes with cataract of children aged >5 years old, with unilateral cataracts were assessed univariately as shown in Figures 43 to 55.

Figure 43: Visual acuity at last examination in 51 eyes with cataract of children with unilateral cataract by time since detection (months)

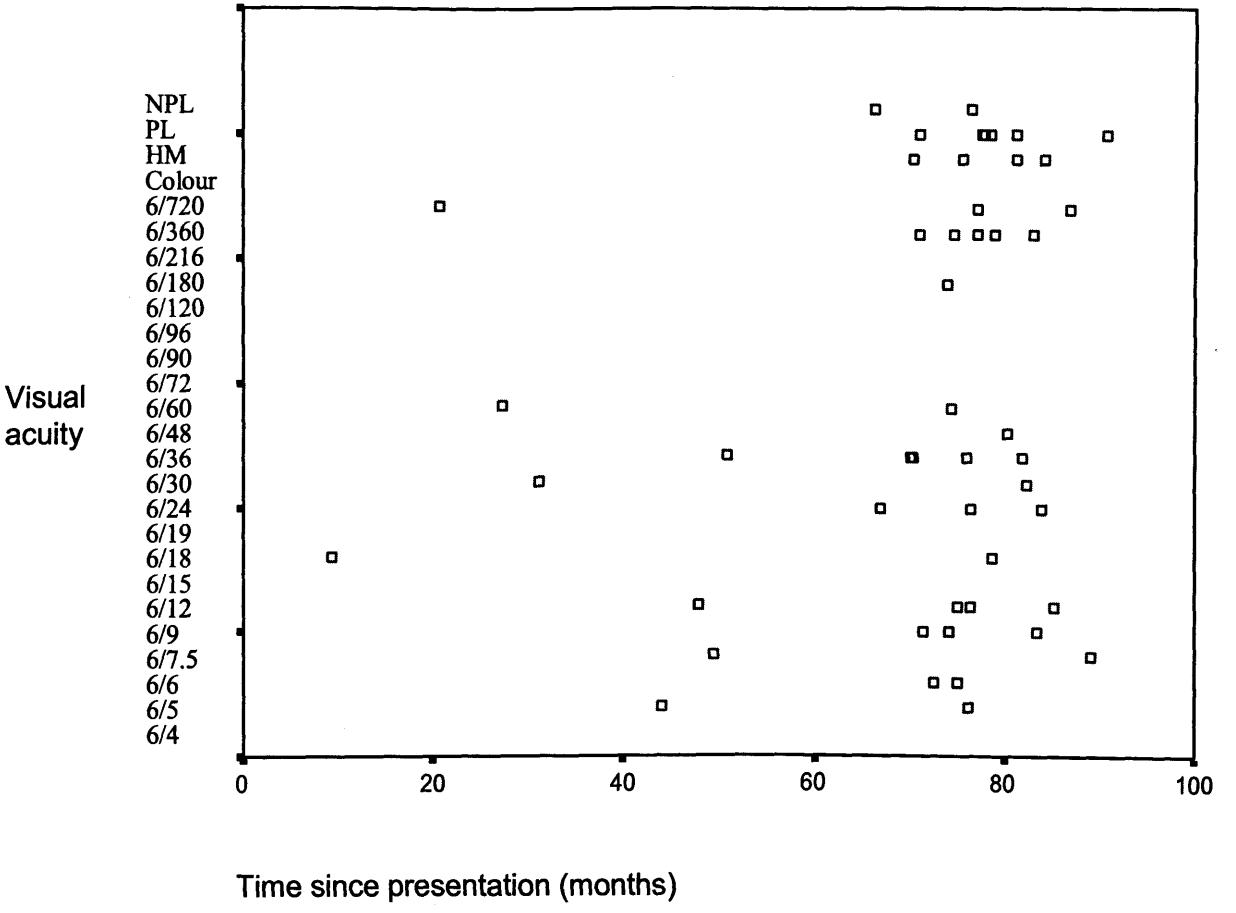


Figure 44: Visual acuity at last examination in 51 eyes with cataract of children with unilateral cataract by age at detection (months)

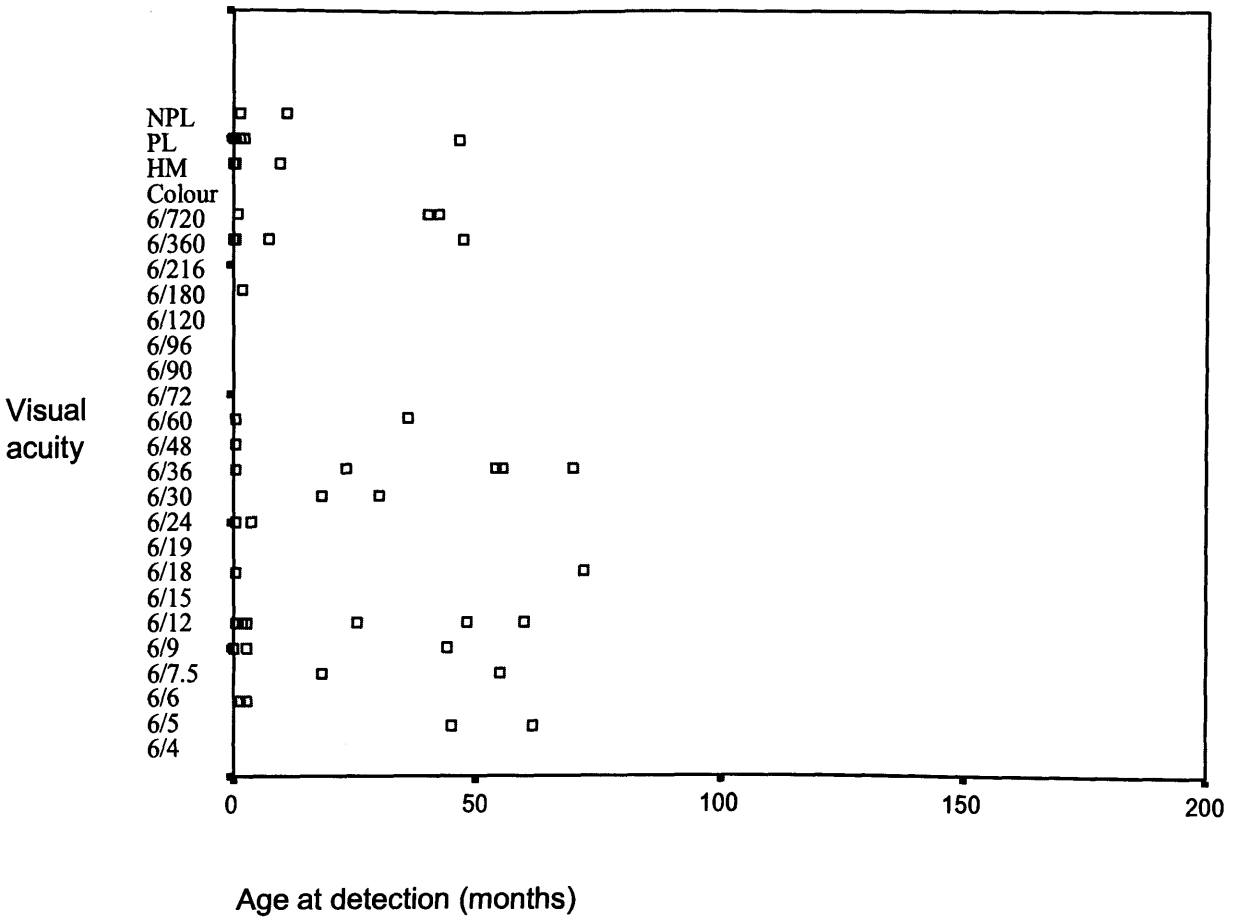


Figure 45: Visual acuity at last examination in 32 eyes with cataract of children with unilateral cataract by time since cataract surgery (months)

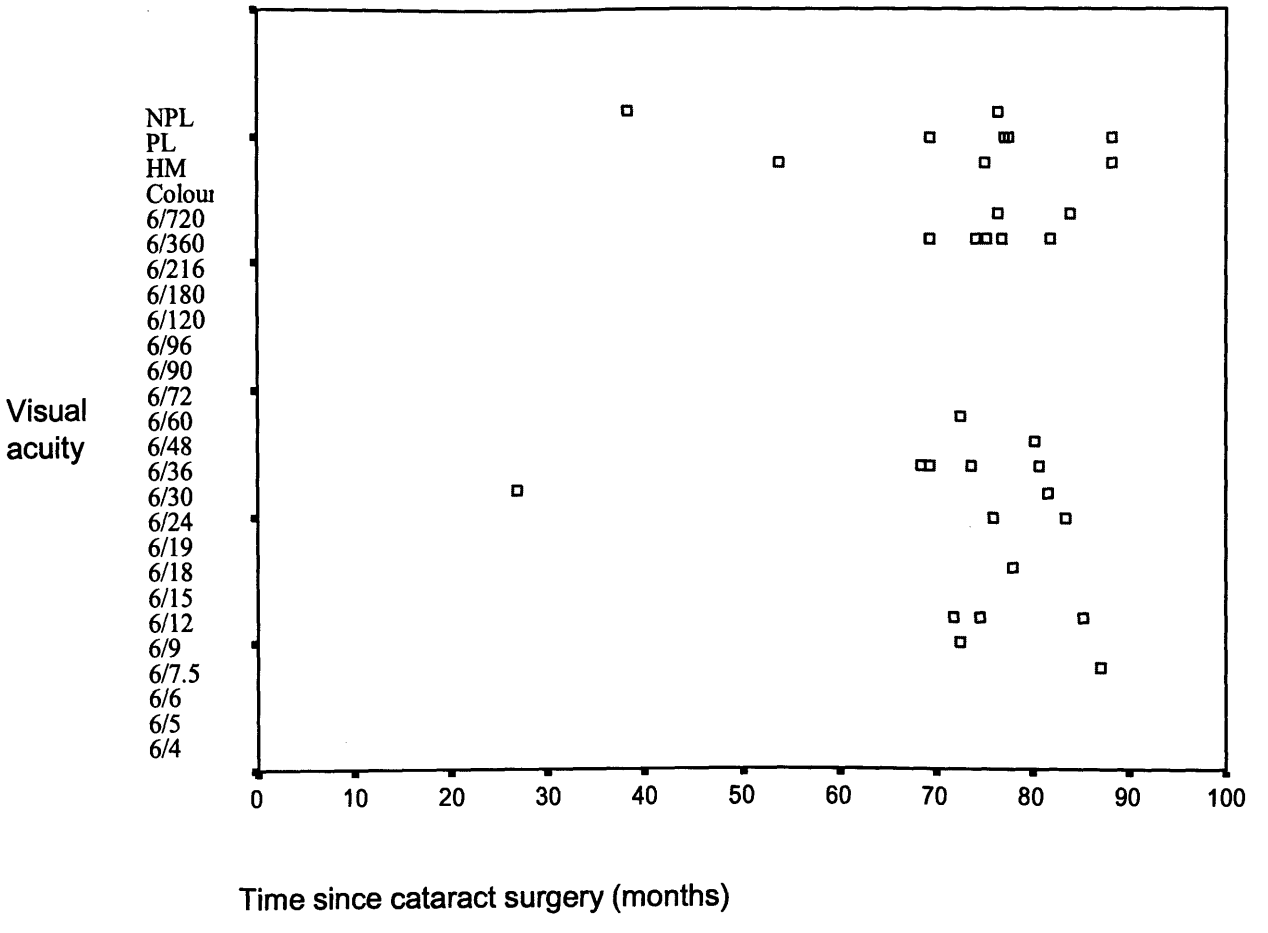


Figure 46: Visual acuity at last examination in 32 eyes with cataract of children with unilateral cataract by age at surgery (months)

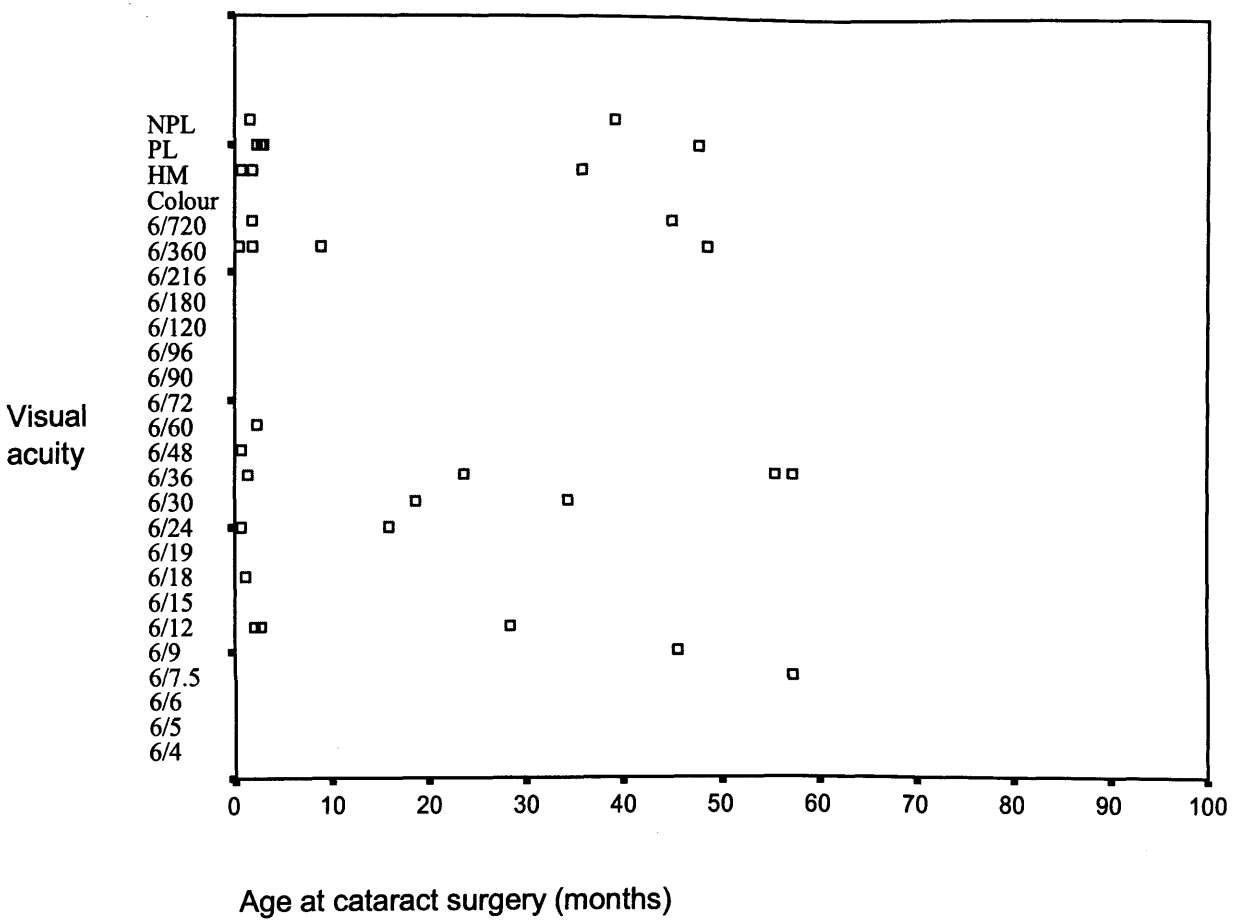


Figure 47: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by final type of correction (N=31)

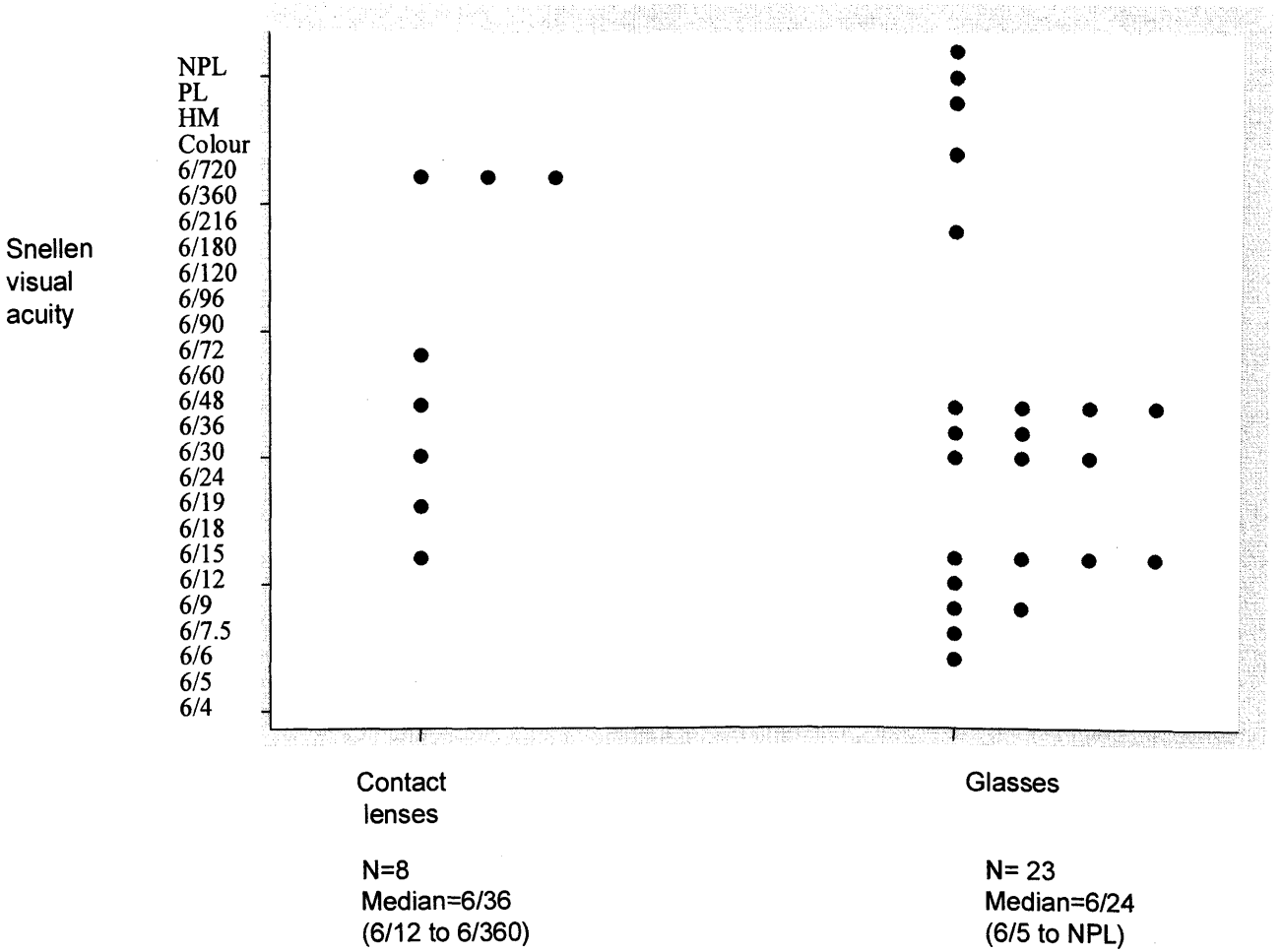


Figure 48: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by type of cataract surgery (N=35)

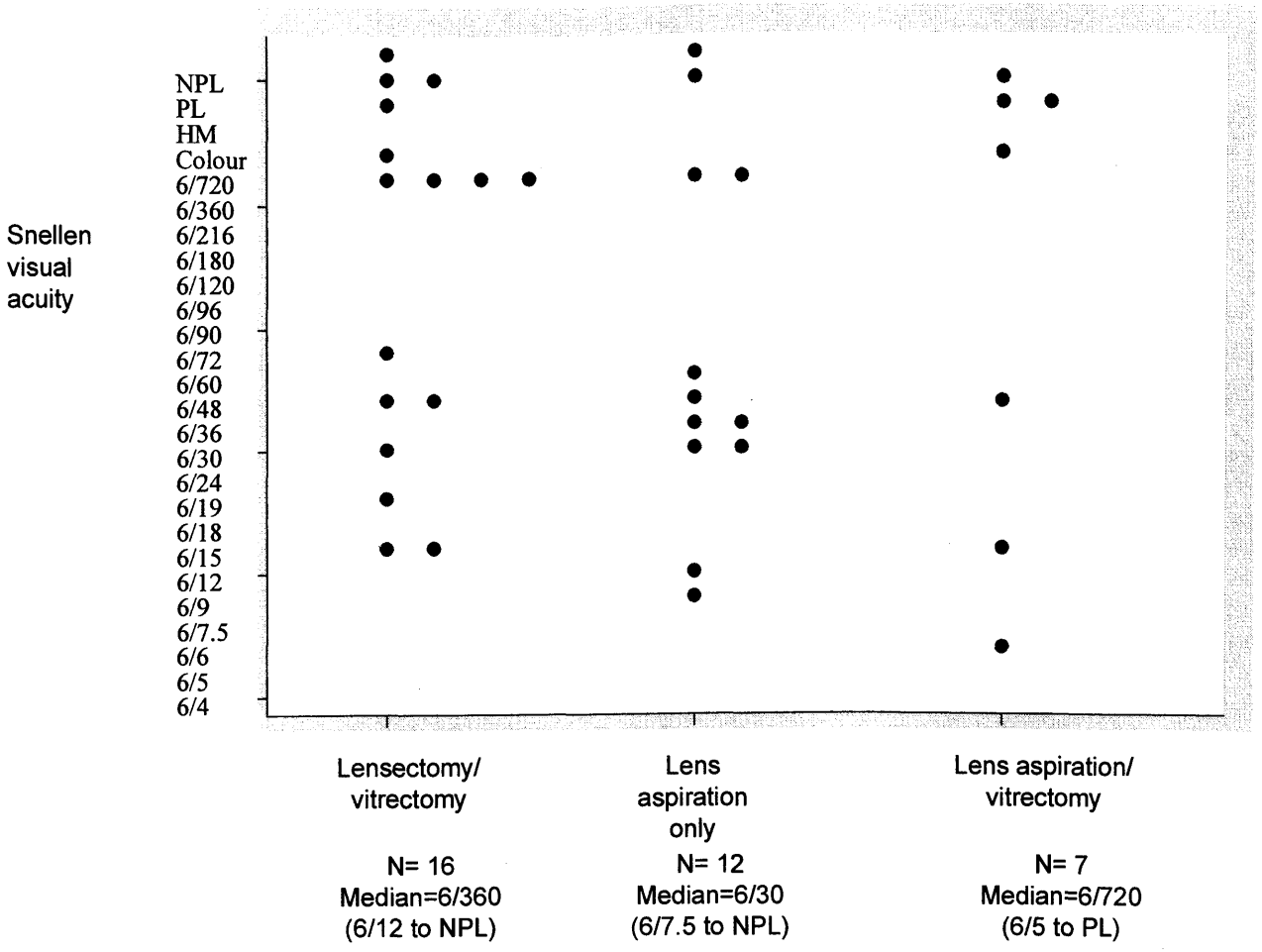


Figure 49: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by primary intraocular lens implantation (N=35)

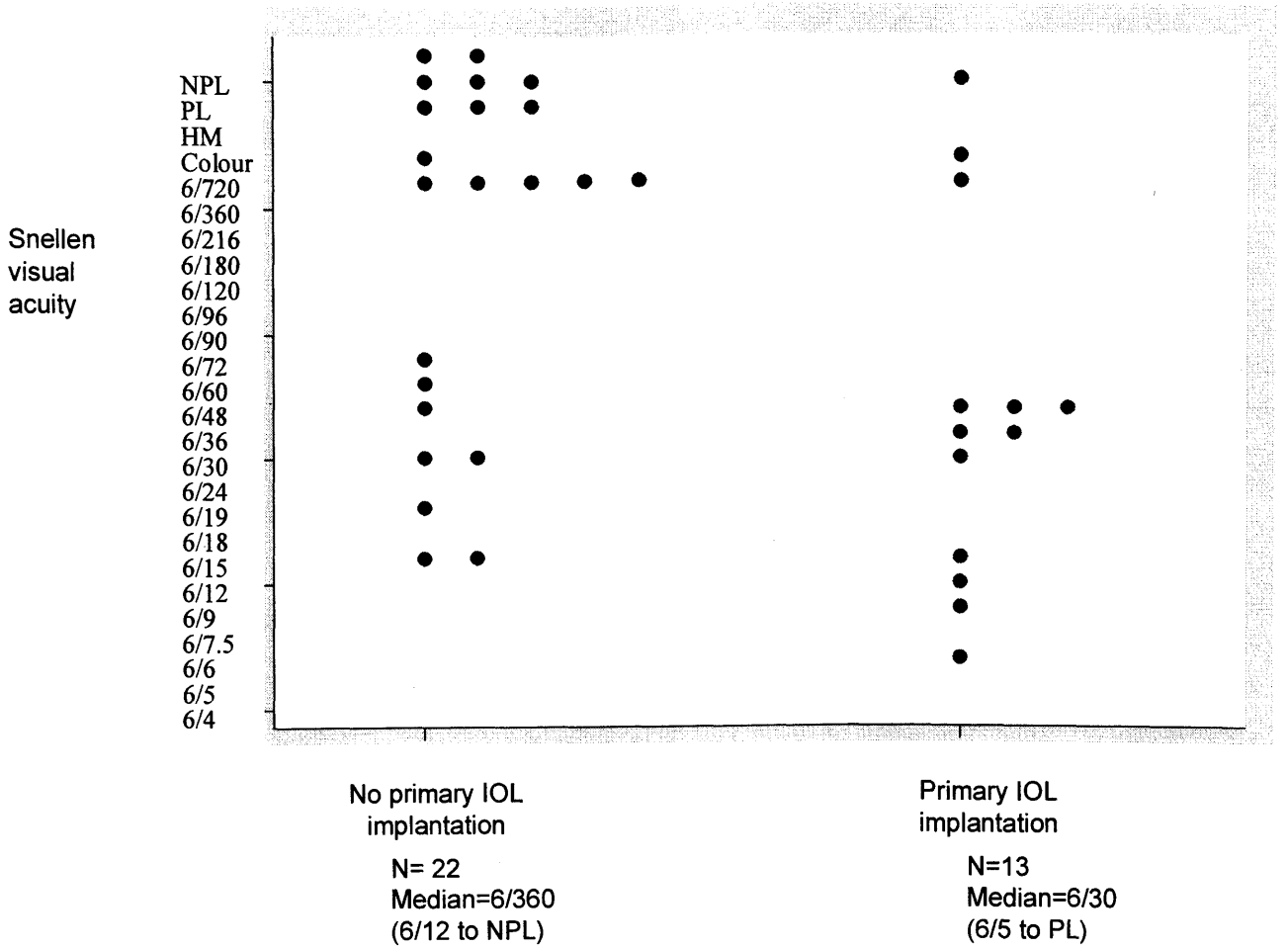


Figure 50: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by concordance with occlusion regime (N=38)

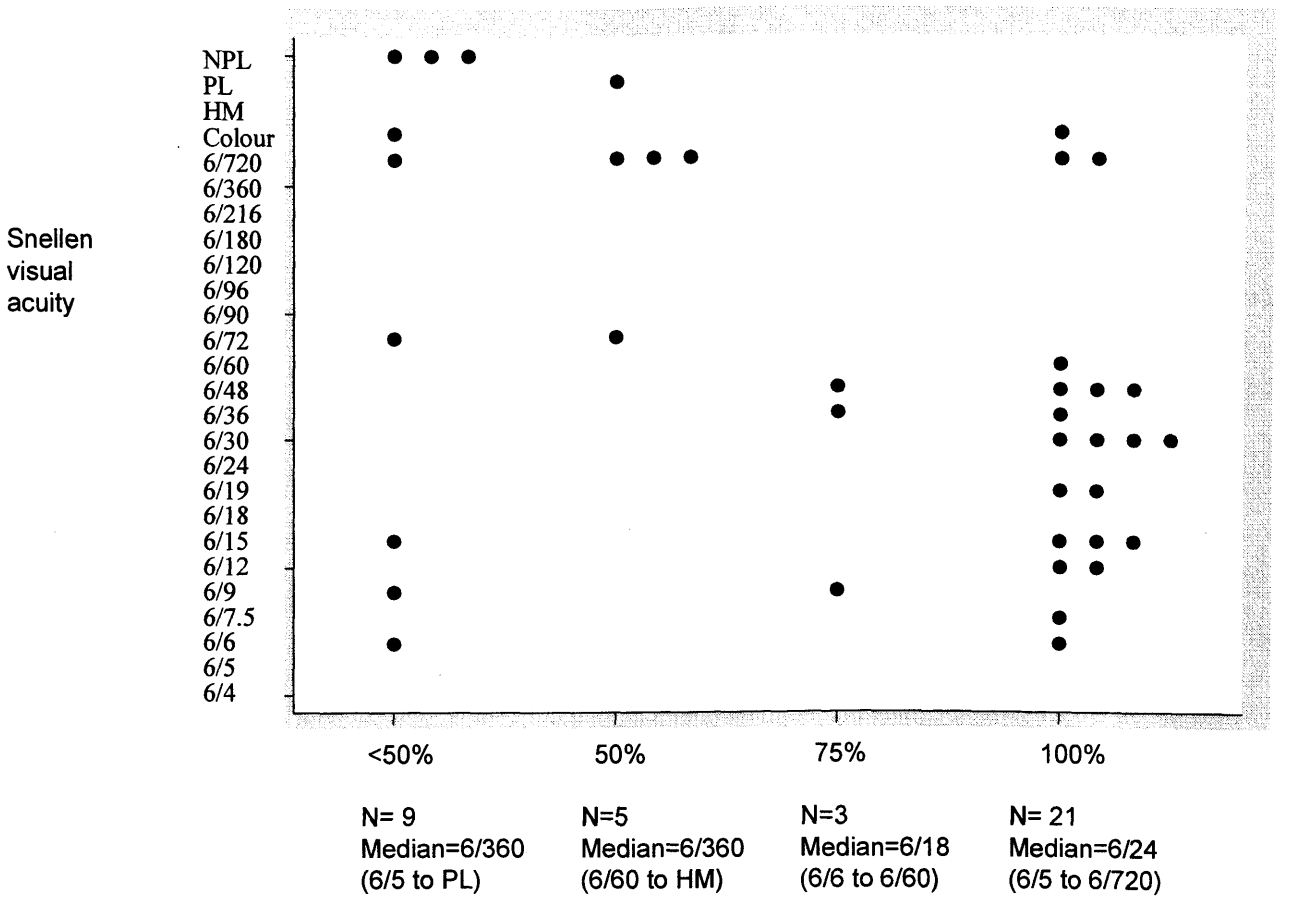


Figure 51: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by aetiological category (N=58)

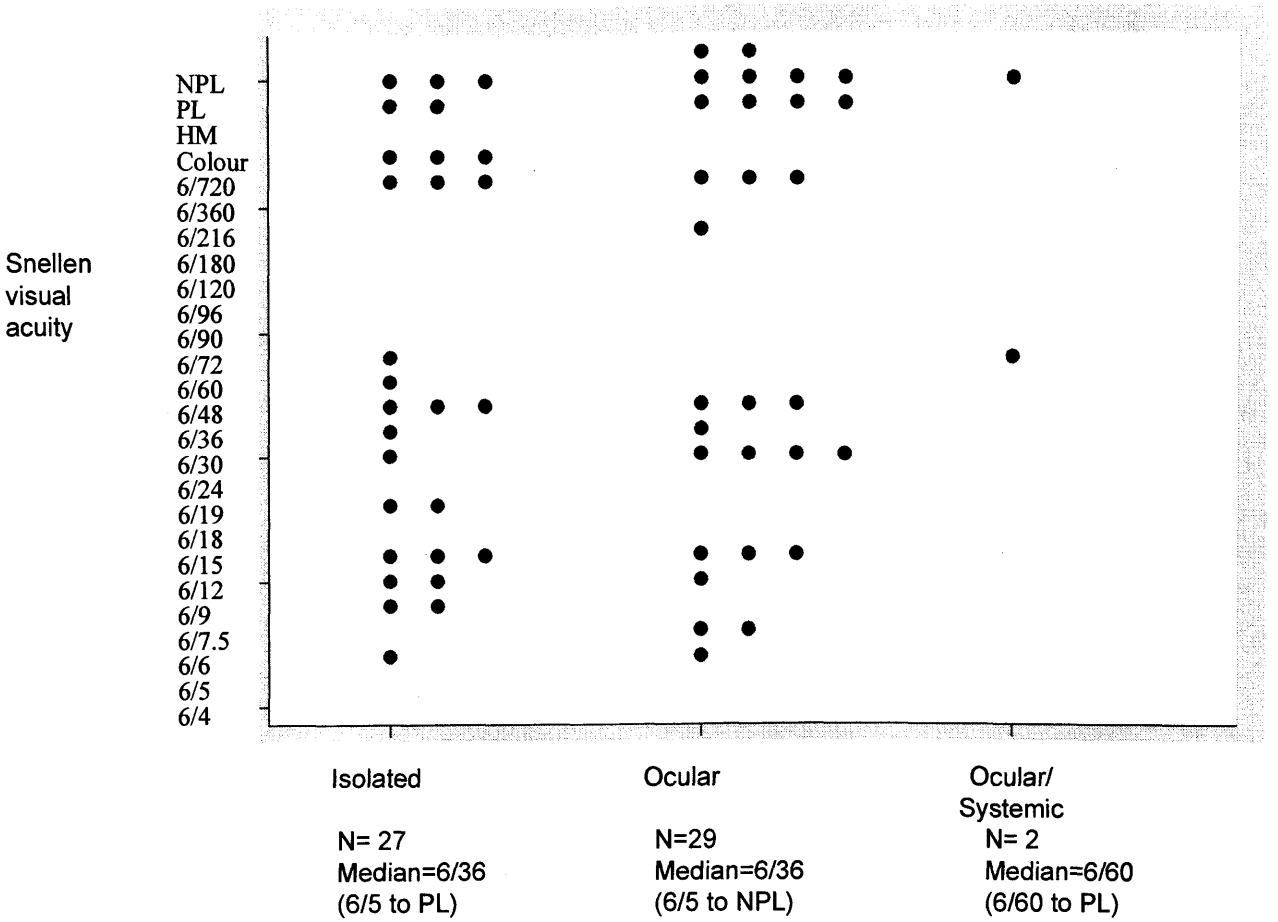


Figure 52: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by presence of other medical disorders (N=58)

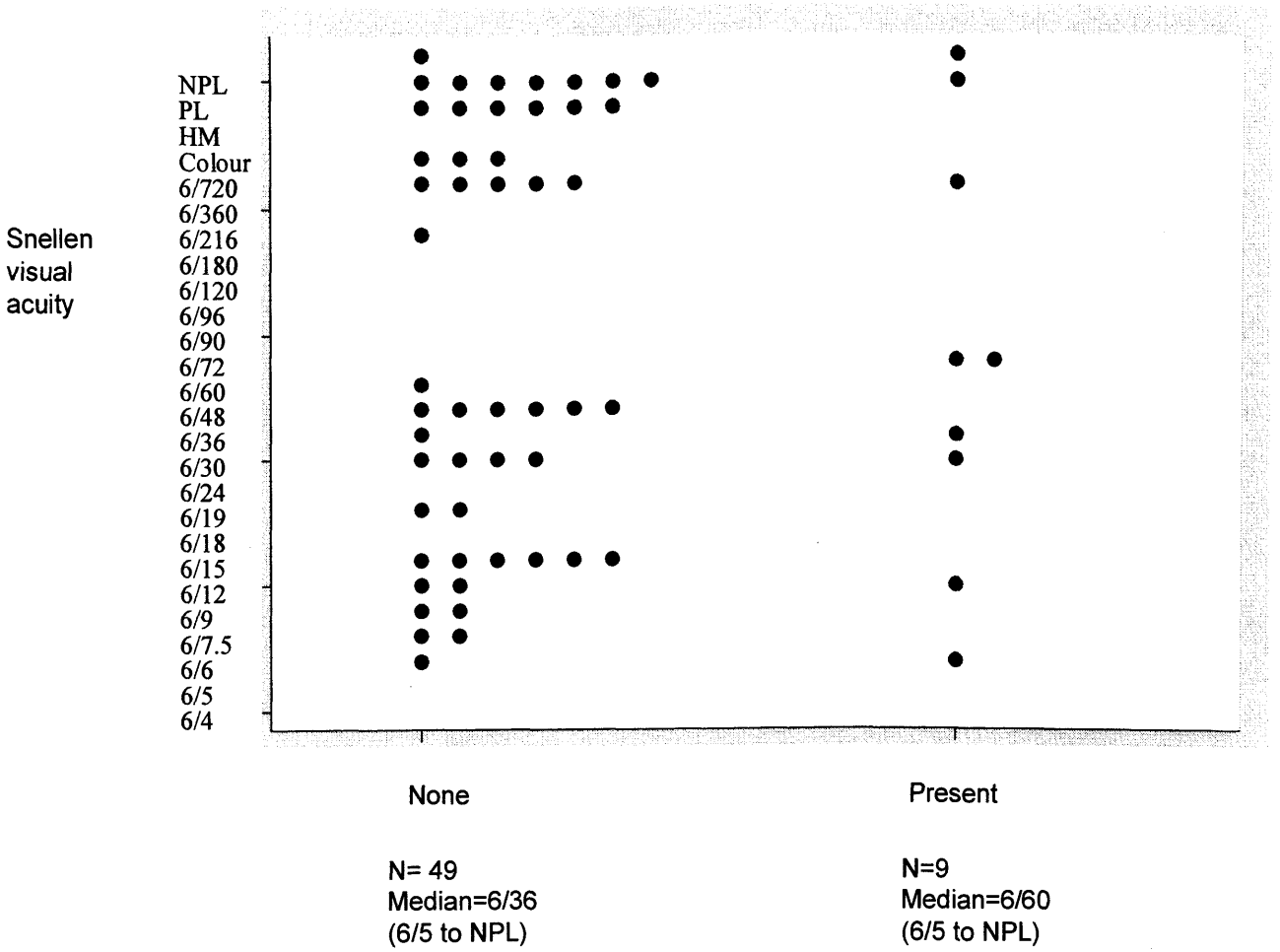


Figure 53: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by severity of cataract at presentation (N=58)

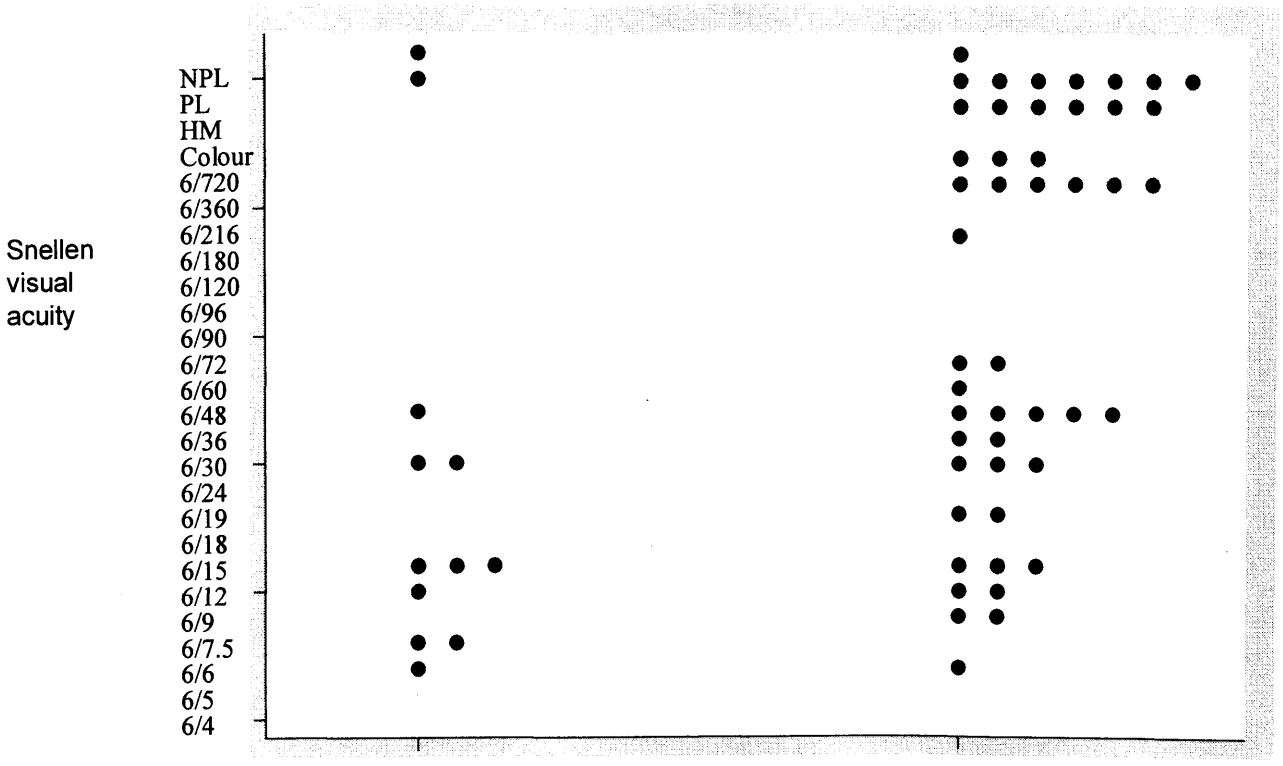


Figure 54: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by occurrence of any postoperative sight threatening complications (N=35)

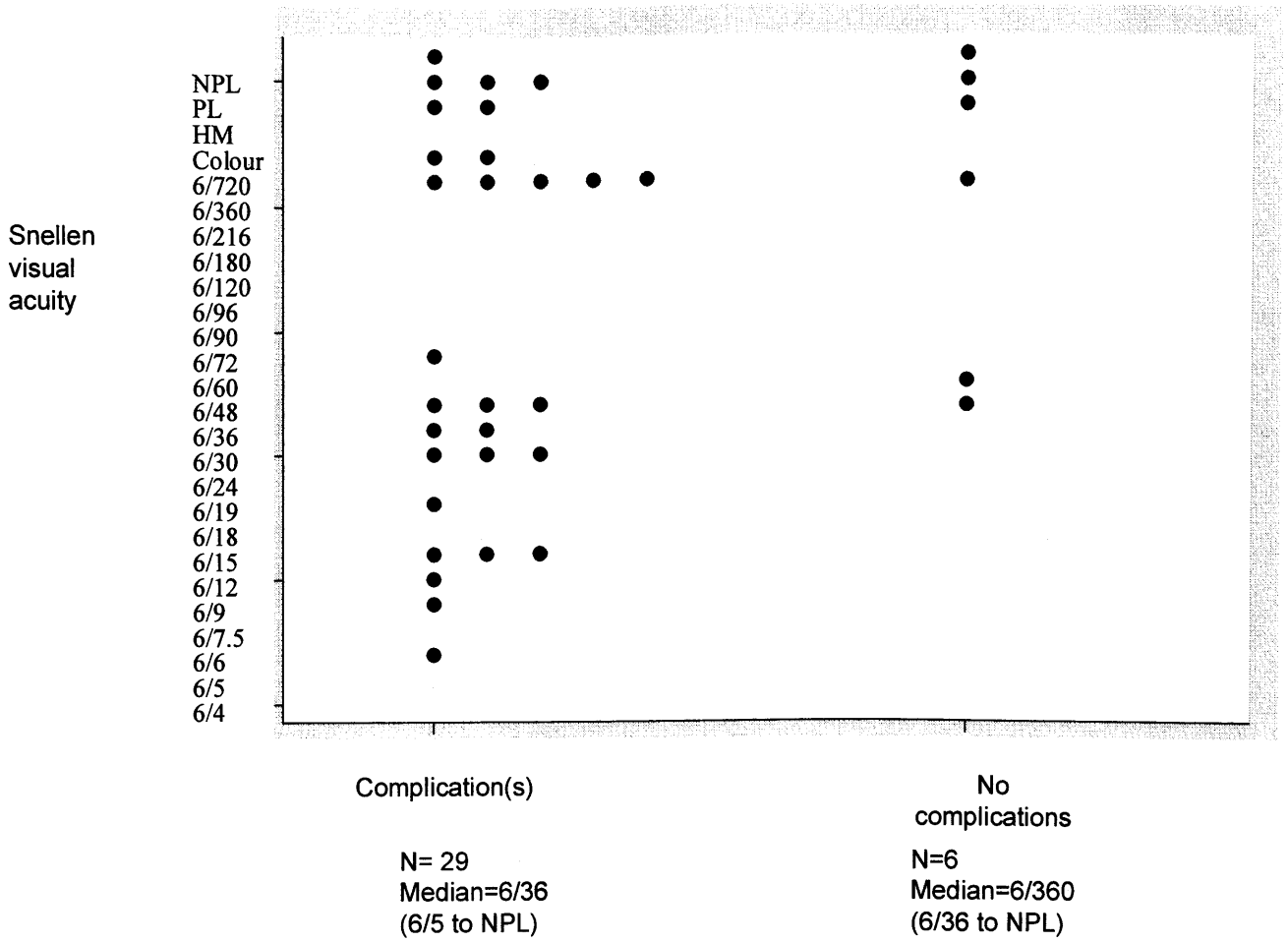
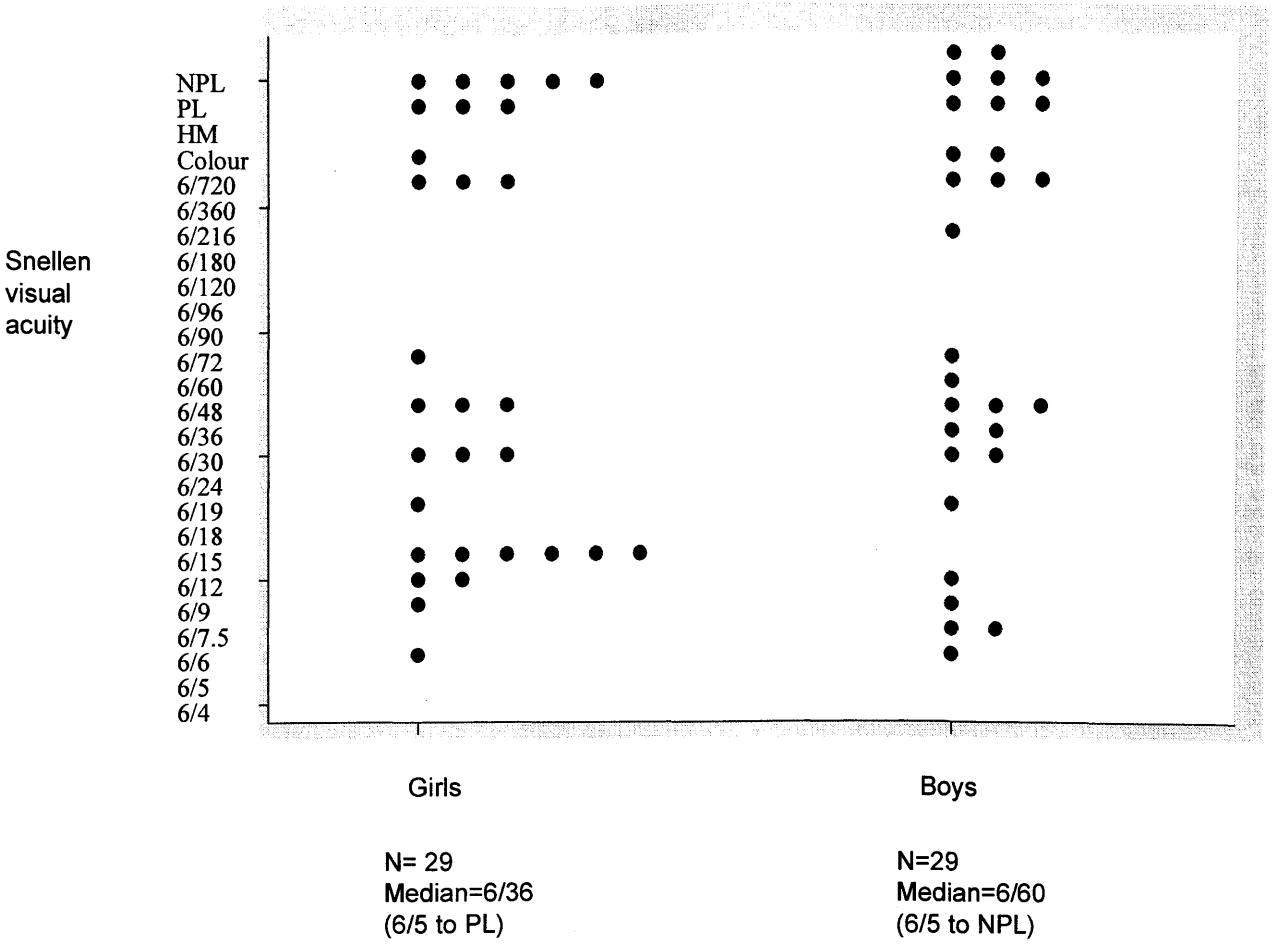


Figure 55: Distribution of visual acuity of eyes with cataract of children with unilateral cataract at final examination by gender (N=58)



10.7 Appendix 7

Factors of interest in relation to the development of postoperative open angle glaucoma are shown in Figures 57-64.

Figure 57: Distribution of glaucoma cases by age at detection

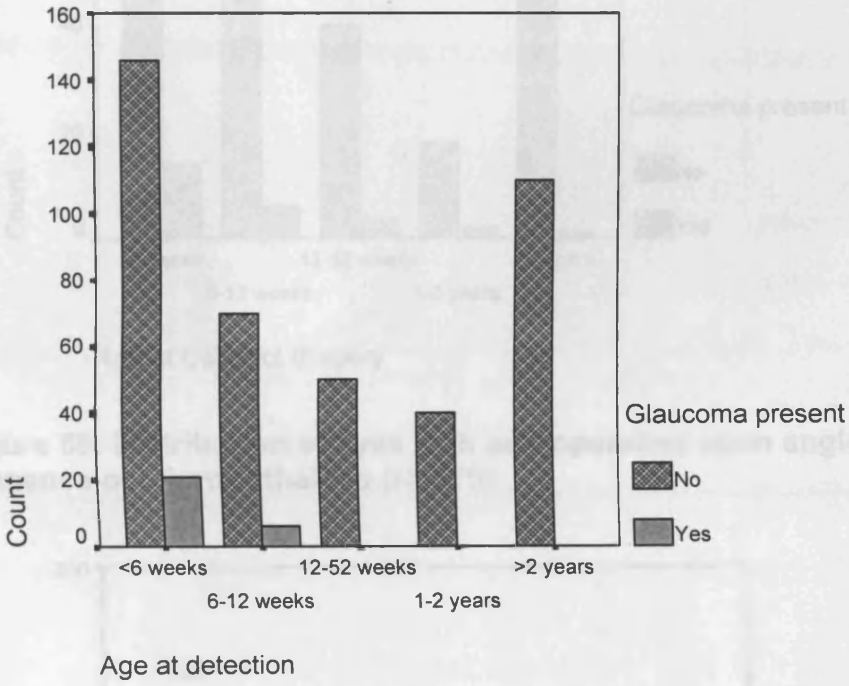


Figure 58: Distribution of glaucoma cases by age at cataract surgery

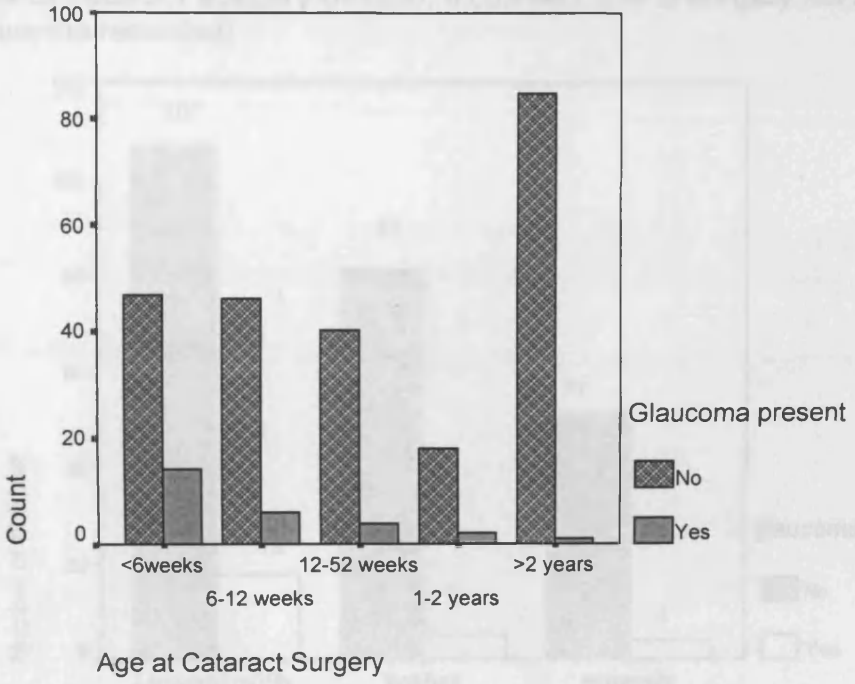


Figure 59: Distribution of eyes with postoperative open angle glaucoma by presence of microphthalmia (N=275)

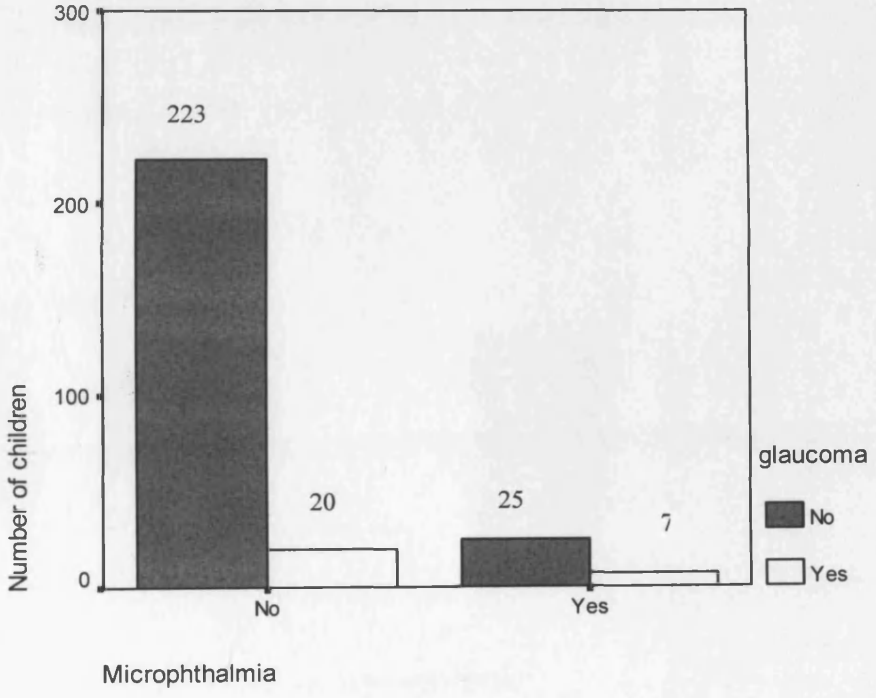


Figure 60: Distribution of eyes with postoperative open angle glaucoma by type of cataract surgery (N=266, 9 children type of surgery not known and no glaucoma recorded)

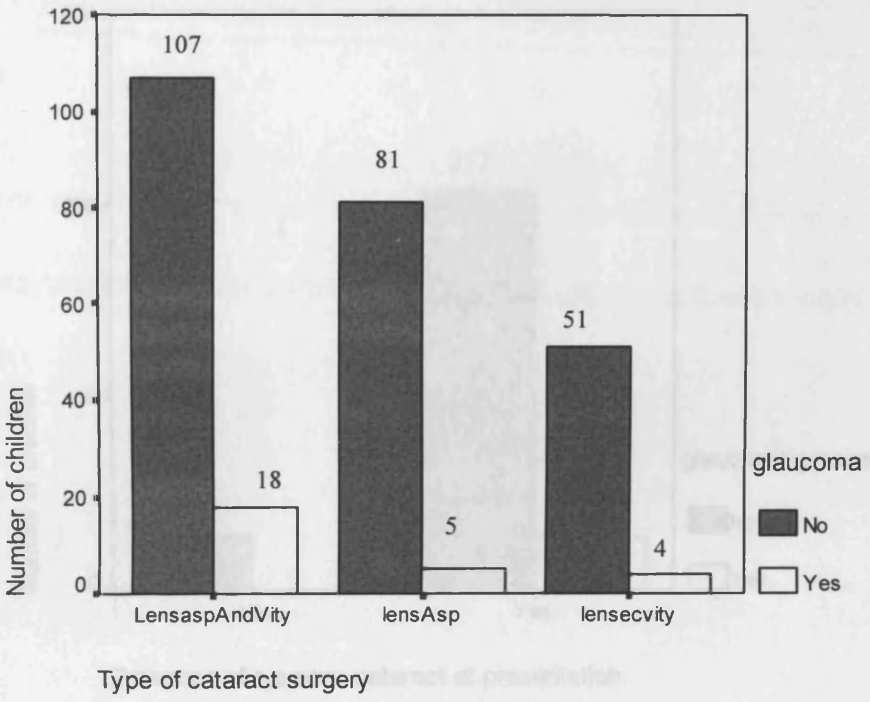


Figure 61: Distribution of eyes with postoperative open angle glaucoma by severity of cataract at presentation (N=273, 2 children severe of cataract at presentation not known, no glaucoma recorded)

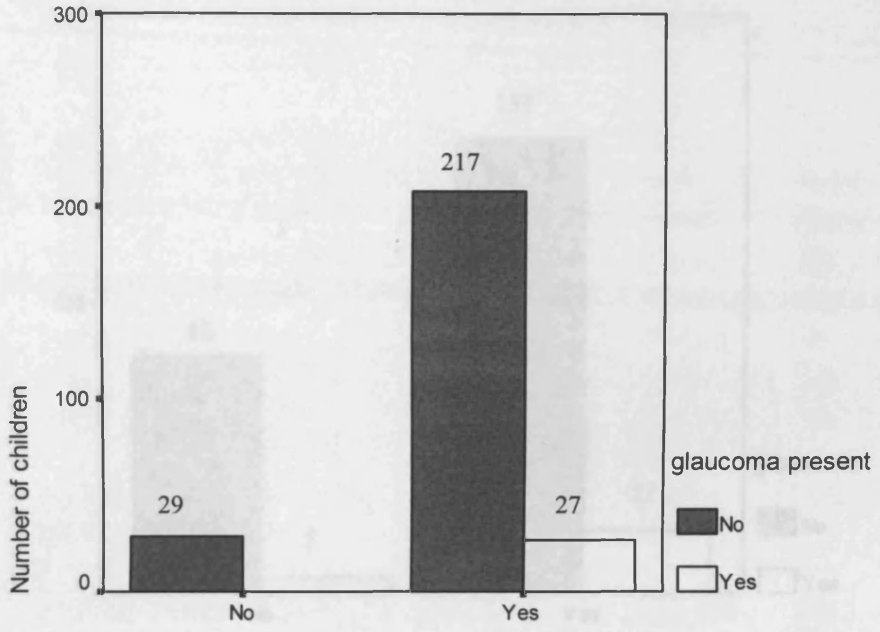


Figure 62: Distribution of eyes with postoperative open angle glaucoma by primary intraocular lens implantation (N=275)

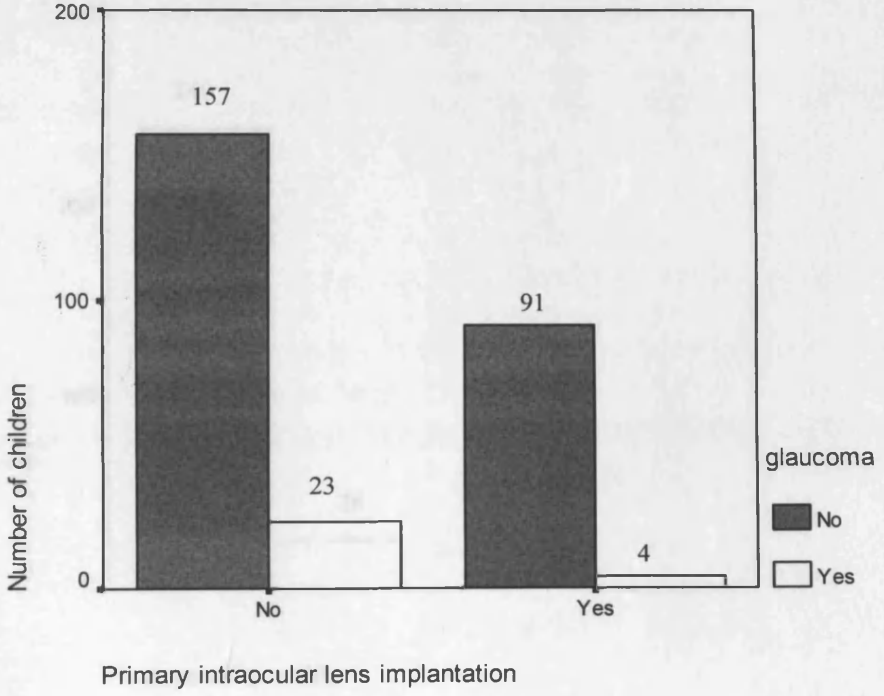


Figure 63: Distribution of eyes with postoperative open angle glaucoma by primary vitrectomy (N=275, 9 children type of surgery unknown, no glaucoma recorded)

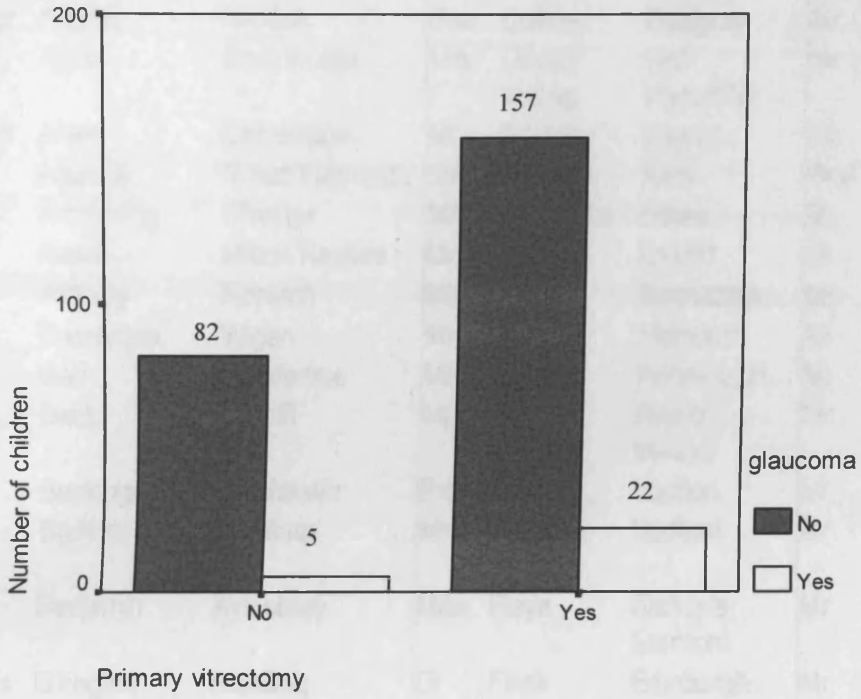
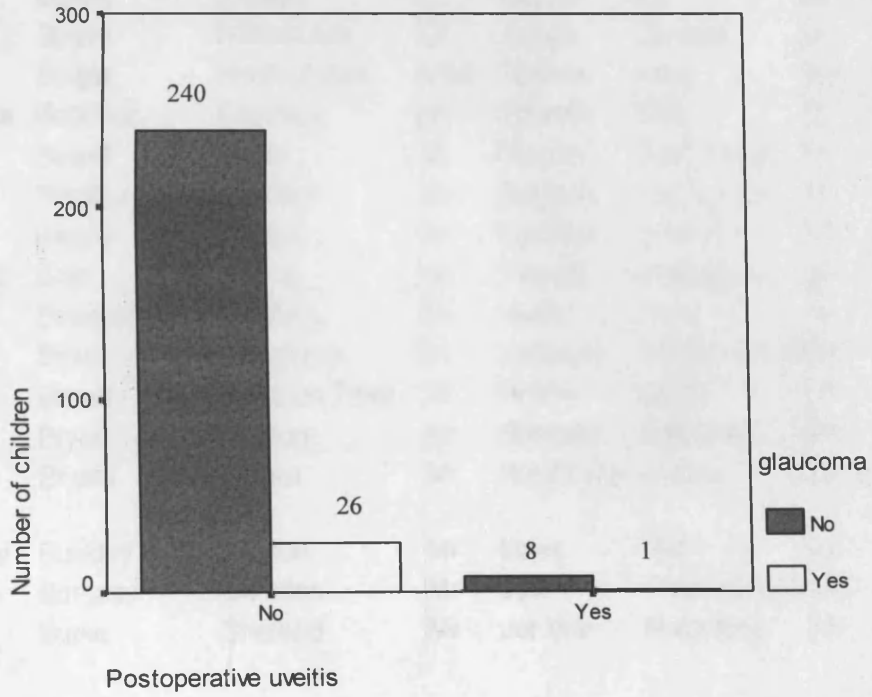


Figure 64: Distribution of eyes with postoperative open angle glaucoma by presence of significant postoperative uveitis (N=275)



10.8 Appendix 8

Members of the British Congenital Cataract Interest Group

Mr	Abdel-Khalek	Boston	Mr	Dodd	Manchester	Mr	McConnell	Kent
Mr	Aclimandos	London	Mr	Doran	Leeds	Mr	McGinnity	Belfast
Miss	Adams	London	Prof	Dutton	Glasgow	Mr	McLeod	Brighton
Mr	Aftab	Scunthorpe	Mrs	Duvall-Young	High Wycombe	Mr	Mishra	Notts
Miss	Allen	Cambridge	Mr	Edelston	Ipswich	Mr	Mohamad	Chesterfield
Mr	Amanat	Great Yarmouth	Mr	Edwards	Kent	Prof	Moore	London
Mr	Armstrong	Chester	Mr	El-Kasaby	Essex	Mr	Moriarty	Cheshire
Mr	Assaf	Milton Keynes	Mr	Elston	Oxford	Dr	Morrice	Stirling
Mr	Astbury	Norwich	Miss	Enoch	Barnstable	Mr	Morris	Southampton
Mr	Bannerjee	Wigan	Mr	Evans	Plymouth	Mr	Munton	Kent
Dr	Barr	Dumferline	Mr	Evans	Portsmouth	Mr	Neugebauer	Cheshire
Ms	Beck	Cardiff	Mr	Fahy	Rep of Ireland	Mr	Newman	Liverpool
Mr	Beckingsale	Colchester	Prof	Fielder	London	Mr	Nischal	London
Mr	Bedford	Dumfries	Mr	Fisher	Bedford	Mr	Nolan	Republic of Ireland
Mr	Benjamin	Aylesbury	Miss	Flaye	Bishop's Stortford	Mr	O'Connor	Republic of Ireland
Miss	Billington	Reading	Dr	Fleck	Edinburgh	Mr	O'Keefe	Republic of Ireland
Miss	Blamires	Leicester	Miss	Frank	Poole	Miss	Ohri	London
Mr	Bloom	London	Dr	Gaskell	Ayr	Mr	Perry	Kidderminster
Mr	Boase	Portsmouth	Dr	George	Dundee	Mr	Phillips	Wirral
Mr	Bolger	Hertfordshire	Miss	Gibbens	Kent	Mrs	Pieris	Bedford
Miss	Boodhoo	Chertsey	Mr	Greaves	Kent	Dr	Power	Dumfries
Mr	Bowell	Dublin	Mr	Gregory	East Sussex	Mr	Price	Cheltenham
Mr	Bradbury	Bradford	Mr	Gregson	Nottingham	Mr	Quinn	Devon
Mr	Brazier	London	Mr	Hardman	Ipswich	Mr	Qureshi	Rochdale
Prof	Bron	Oxford	Mr	Haworth	Nottingham	Mr	Rahman	Boston
Mr	Brosnahan	Sheffield	Mr	Heravi	Kent	Mr	Rennie	Aberdeen
Mr	Brown	Shropshire	Mr	Hodgkins	Southampton	Mr	Ridgway	Manchester
Mr	Brown	Stoke on Trent	Mr	Holden	Derby	Mr	Roper- Hall	Birmingham
Mr	Bryan	London	Mr	Humphry	Salisbury	Mr	Rosen	Manchester
Mr	Bryars	Belfast	Mr	Hutchinson	Halifax	Miss	Russell-Eggitt	London
Prof	Buckley	London	Mr	Innes	Hull	Mr	Shun Shin	Wolverhampton
Ms	Burgess	Swindon	Mr	Jalili	Peterborough	Mr	Simcock	Exeter
Mr	Burke	Sheffield	Mr	Jenkins	Maidstone	Mr	Simmons	Leeds

Ms	Butler	Birmingham
Mr	Calver	London
Mr	Casswell	Brighton
Mr	Chandna	Liverpool
Mr	Church	Aberdeen
Mr	Clarke	Middlesborough
Mr	Clarke	Newcastle upon Tyne
Dr	Coffey	Republic of Ireland
Mr	Cole	Devon
Mr	Condon	Chertsey
Mr	Corridan	Wolverhampton
Mr	Dang	Darlington
Mr	Darvell	Kent
Mr	Das	Worcestershire
Mr	Davies	Norwich
Mr	Daya	West Sussex
Mr	De Cock	Margate
Mr	Dees	Darlington
Dr	Johnson	Gloucester
Mr	Kaushik	Wrexham
Mrs	Kayali	London
Mr	Keightley	Basingstoke
Prof	Khaw	London
Mr	Kinnear	London
Mr	Kotta	Grimsby
Mr	Kumar	Cornwall
Dr	Lavy	Glasgow
Mr	Laws	Swansea
Miss	Leitch	Surrey
Mr	Liu	Brighton
Mr	Lloyd	Manchester
Ms	MacEwen	Dundee
Mr	Macfarlane	Kent
Mr	Mackintosh	Cheltenham
Mr	Mandal	S Yorkshire
Mr	Markham	Bristol
Mr	Tappin	Surrey
Mr	Taylor	York
Prof	Taylor	London
Dr	Thaller	Plymouth
Mr	Thoung	Essex
Mr	Tormey	Republic of Ireland
Mr	Tuft	London
Mr	Tutton	Chester
Mr	Twomey	Somerset
Mr	Verghese	West Cumberland
Ms	Vickers	Brighton
Mr	Vijaykumar	Blackburn
Mr	Vivian	Bury St Edmunds
Miss	Williams	Bristol
Mr	Willshaw	Birmingham
Mr	Woodruff	Leicester
Mr	Wright	Burnley
Mr	Young	Republic of Ireland
Mr	Zaidi	S Yorkshire